

PEDIATRICS

Handwritten Note

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Name: _____

Subject: _____

PEDIATRICS



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PEDIATRICS

Neonatal - First 4 wk. of life.

↳ Early - 7 days, cause of death - Prematurity
Late - 7-28 days, - Sepsis.

Term - 37-42 wk gestation

Preterm - < 37 wk

Post term - > 42 wk

Normal wt \rightarrow 2.5-4 kg

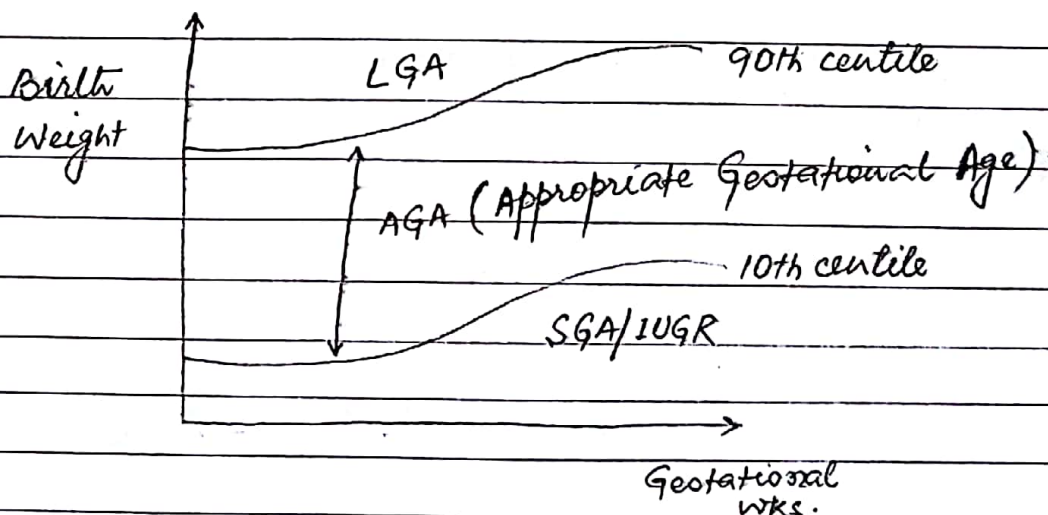
LBW < 2.5 kg

VLBW < 1.5 kg

ELBW < 1 kg

Macrosomia > 4 kg.

Lubchencho chart:



Constitutional:

- LGA (large Gestational Age)
- Short stature.
- Delayed puberty.



- All term baby has 6 fontanelle:
 - Anterior → ~~diamond shaped~~
 - Posterior
 - 2 sphenoid
 - 2 Mastoid.

Posterior fontanel - Closes at birth.
open in 3% baby.

Anterior fontanel - diamond shaped.

2.5 X 2.5 cms.

At level

Pulsatile

Closes at 18-24 months of life.

Craniosynostosis: ^{all}
Early closure of fontanel.

Complication:

- Microcephaly
- ICT ↑ → optic atrophy.
- cosmetic.

R - Craniectomy

Syndromes associated - Apert
Crouzon } AD
Pfeiffer }
Carpenter - AR

Delayed closure of fontanel:

- Rickets
- Down's
- Hypothyroidism
- Hypophosphatasia
- Cleido cranial dysostosis.



- Q. 2 wks baby, Hypotonia, hypothermia, umbilical hernia, constipation, physiological jaundice prolonged.
↳ Congenital hypothyroidism
(humeral head - Epiphyseal dysgenesis)
(Abn epiphysis of bone)

Neonatal Screening:

- TSH ; T_4
 - Delayed rise of TSH
 - TBG deficiency (TSH \uparrow ; $T_4 \downarrow$)
 - Best time : after 48hrs (72hrs)

M/c/c of congenital hypothyroidism - 85% agenesis/
dysgenesis of thyroid gland.

Congenital hypothyroidism:

Prevalance = 1:2000

Girls > Boys.

Neonatal screening : Continue

- TSH ; T_4 • Cold temp^r
- Physiological TSH surge 48hrs.
 - APP > 48hrs to 6days.
 - In OP Ghai \rightarrow 3-5days



\rightarrow Sample obtained \rightarrow Heel prick.
Safe area \rightarrow Side prick.

- pt in shock, venous access can't get in 60sec.
- M/c ^{other} site - Near Tibial tuberosity.
Upper end of tibia
Lower end of femur.
- In shock \rightarrow i.v. fluids \Rightarrow 20ml/kg bolus NS.



Most easily assessable venous route - Umbilical Vein.

Phenylketonuria: Deficiency of Phenylalanine hydroxylase.

Phenylalanine
PKU (-) \nrightarrow Phenylalanine hydroxylase.

~~Phenylalanine~~

Tyrosine
 \downarrow Tyrosinase

DOPA

\downarrow

Dopa quinone
 \downarrow

Melanin

\uparrow Phenylalanine \rightarrow Toxic to Brain \rightarrow so in PKU High Phenylalanine

- Child Mental Retardation.

20 mg/dl

- Developmental delay.
- Exaggerated Reflexes
- Microcephaly.

Rx - PKU

- Supplement Tyrosine
- Restrict Phenylalanine
- Now Tyrosine essential.
- Aim: Serum Phenylalanine < 6 mg/dl.
- Lifelong.

Q. About PKU Rx, first step -

- A) Stop the substrate of the enzyme
- B) Supplement the enzyme.
- C) Reduce the substrate of the enzyme
- D) Provide deficient proteins.



Maple Syrup Urine disease (MSUD):

- Deficiency of α -keto acid branched chain dehydrogenase.
- High Valine : Leucine : Isoleucine blood & CSF.
- Coma
- Severe acidosis.
- We do dialysis - To remove Valine : Lev : Isoleucine.

Neonatal Screening : Continue

- Tandem mass
 - Spectrophotometry \rightarrow metabolic
- Cystic fibrosis
- Congenital adrenal hyperplasia
- G6PD deficiency.
- Biotin deficiency.

ANTHROPOMETRY : TERM BABY

Length - 50 cm

Head circumference - 35 cm

HC > CC but not more than 3 cm.

- HC > CC ($>> 3$ cm) \rightarrow Congenital hydrocephalus.
 \rightarrow Asymmetric IUGR baby.

CC = HC \rightarrow at 9-12 months

By 1 yr ; CC $>>>$ HC

Upper Segment : Lower segment (US : LS)

New born = 1.7 : 1

At 10 yrs = 1 : 1

Adults = 0.9 : 1

Achondroplasia : Short limb dwarf (US/LS \uparrow)

Hypochondroplasia : US/LS \uparrow (disproportionate short stature)



Meconium:

95% pass meconium at 24 hrs.
99% " " " 48hrs.

Cause of delayed passage meconium

- Imperforate anus
- Hirschsprung / aganglionic
↳ Rectum biopsy.
- Meconium ileus → Cystic fibrosis.
↳ Small intestinal obstruction.

Q 48hrs baby has not passed meconium. Next Ix

- A) CFTR gene test
- B) Sweat chloride
- C) Manometry
- D) Lower GI contrast study.
↳ Δ Hirschsprung
Treat meconium ileus.

Delayed Urine:

- B/L Renal agenesis
↳ Maternal oligohydramnios.

Post. Urethral valve ^{also cause} ↓
B/L pulm. hypoplasia
⇓
POTTER'S Sequence.

Potter's face - Nose pinched in

Retrognathia

Micrognathia

Bag & mask → Pneumothorax.



Q If a baby has not passed urine in 1st 48hrs, Next Ix
→ USG

Q 3 days old, c/o - Weak dribbling urine stream.

O/E - palpable distended bladder.

Δ → Posterior Urethral valve.

IOC : MCU (Micturating Cysto Urethrography).

Rx : Cystoscopic Fulguration

SGA/IUGR :

Complication : TORCH infection / Chromosomal disorders.

Symptom - Asphyxia → causes MAS



(Meconium Aspirated Syndrome)

PPHN (Persistent pulm. HTN of New born).

Severe IUGR - Pulmonary hemorrhage

Limited stores - Hypoglycemia, HypoCa; HypoMg.

- Polycythemia

↳ bcoz in IUGR erythropoietin is very sensitive to hypoxia.

- Neutropenia

- Thrombocytopenia.

Q Full term small for date babies are more disposed to -

a) HyperCa

b) CNS infection

c) PDA

} common in preterm.

↳ Hypoglycemia



	Symmetrical IUGR	Asymmetrical IUGR
Cause →	Chromosomal/ Toxins	Maternal Complications
Cell no.	↓	(N)
Cell size	↓	↓
Brain	↓	(N) spared. HC > CE > 3cm Brain/liver ↑

PONDERAL'S INDEX:

$$\frac{\text{Wt (gm)}}{\text{Length (cm)}^3} \times 100$$

< 2 → Asymmetrical IUGR

> 2 → AGA/Symmetrical IUGR

Normal Neonatal phenomenon:

Milia: Distended Sebaceous gland on face & nose.

Erythema Toxicum: Erythema on face & trunk
2-3 days of life.

Stork bites: Pinkish gray capillary hemangioma
on back & buttocks.

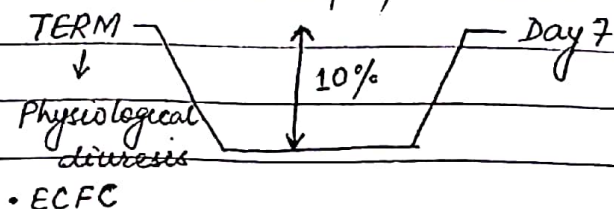
Epstein Pearl: Epithelial inclusion cyst on palate
& prepuce.

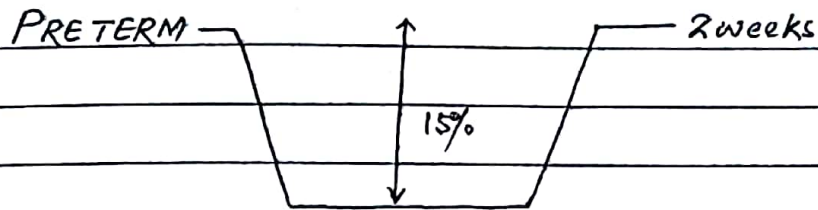
Natal teeth (Pre deciduous teeth): lower incisor position.

~~Wid~~ Withdrawl vaginal bleeding: On 5th-7th day.

Acrocyanosis (Peripheral Cyanosis): Limbs cyanosed
Lips pink.

Birth wt. = 3000 gm
Day 4 = 2700 gm
Day 7 = 3000 gm





IUGR → do not lose wt (ECF compact)
↳ Wt. stable for 1-2 days then wt. gain.

Q.

Not normal in a newborn?

- A) Proteinuria
 - B) Glucosuria
 - C) 1-2 pus cells/hpf
 - D) Bacteriuria
- } in ELBW

Neonatal Reflexes:

① Moro Reflex:

1st phase - Abduction of shoulder joint
Extension of elbow joint.
opening of fingers

2nd phase - Adduction & flexion.

- Appears 28-32 wks gestation.
- Adduction/complete → 36-38 wks.
- Disappears at around 2-3 months of life.
- Persistence beyond 6 months abnormal.



Cerebral palsy. ^Q

Q - Asymmetric moro's

- Brachial plexus injury.
- # clavicle
- # Humerus
- Hemiplegia.



- # Early hand preference is always abnormal
- Hemiplegia at other side.
 - 95% cases → Hemiplegia at Rt. side.

Exaggerated Moros → HIE-1

ATNR (Asymmetric Tonic Neck Reflex):

- Side of face - Extended
- Side of occiput - Flexed.
- Onset - 35 wks
- Fully developed - 1 month
- Duration - 6-7 months
- Do not ~~roll~~ roll (Rolling start when this reflex disappears)
- Disappears - 6 months

STNR (Symmetric tonic Neck reflex):

→ Neck extended
→ Tone ↑ UL
↓ LL

Neck flexed
↓

Tone ↓ in all limbs.

- Not present at birth.
- Appears 4-6 months of life.
- Disappears 8-12 months of life.
- Child start to crawl when this reflex disappears.

Parachute reflex:

- Not at birth
- Appears 6-7 months of life.



- Well developed at 10-11 months of life.
- Persists life time.

LANDAU Reflex:

- Appears at 3 months of life
- Disappears at 1 year of life.

On ventral suspension - Spine strengthen /
Straighten.

Child get out of flexion attitude by this reflex.

GRASP Reflex:

- Appears 28 wks of gestation.
- Well developed 32 wks
- Disappears 3 months of life.

Sucking & Rooting reflex:

< 28 wks absent.

At 28 wks - Some sucking bursts.

32 wks - Appear

34 wks - Co-ordination

Q 31 wks; 1500gms; Feed - ?

- A) Enteral → NG tube (Expressed Breast Milk)
- B) Enteral + i.v. fluids
- C) IV fluids.
- D) TPN.

Q 33 wks; 1500gms; Feed - ?

↳ Enteral → Katori / Paladay / Spoon
(Expressed Breast Milk)



AIIMS NICU Protocol 2014 :

- > 34 wks - Breast feed
- 32-34 wks - Katori (Expressed breast milk)
- < 32 wks - NG tube (" " ")
- < 1200 gms - i.v. dextrose + minimal enteral feeds 10-15 ml/kg/day.

↓
Rapid enteral feed can cause - Necrotising Enterocolitis.

Fetal alcohol Syndrome :

- Skin folds at the corner of the eyes
- Low nasal bridge.
- Short nose
- Indistinct philtrum (groove b/w nose & upper lip)
- Small head circumference (microcephaly)
- Small eye opening.
- Flat mid face / midfacial or maxillary hypoplasia.
- Thin upper lip.
- Septal defects ← ASD
VSD.

Q Ass. C ~~Which~~ Fetal alcohol Syndrome except -

- A) Microcephaly
- B) Overgrowth
- C) Flat face
- D) Small palpebral fissure.



PREMATURITY

Respiratory System: RDS (Respiratory distress Syndrome).

Chr. lung disease /

Bronchopulmonary dysplasia

O₂ dependence on
4 wks of life

Rx: Home O₂ therapy.

Prevention: Nasal CPAP.

Vit. A supplementation.

Furosemide

Keep underhydrated in ICU.

CNS: ① Apnea (>20sec); or any period of ass. c central cyanosis & bradycardia).

Q M/c Signs of acute hypoxia in neonates

A) Bradycardia

B) Tachycardia.

Types of Apnea: 3 types

① Central: asphyxia; preterm.

② Obstructive:

all newborn are obligate nose breathers till
4 months of life.

Q Full term newborn

episodes of cyanosis - worsen when feed.

Seems better when crying.

↳ Choanal atresia → B/L, posterior.

③ Mixed (M/c)



Apnea of prematurity:

Risk: \square ≤ 28 wks $\rightarrow 100\%$

Onset: 1-2 days; never > 7 days.

Rx:

1st step: Nasal CPAP

Methylxanthines - Aminophylline (Narrow therapeutic range)

Caffeine citrate (DOC)

\hookrightarrow Wide margin of safety

Q Loading dose of Aminophylline: 5-6 mg/kg.

followed by maintenance dose

1-2 mg/kg every 6-8 hrs.

Caffeine Citrate: loading dose $\rightarrow 20$ mg/kg

Maintenance dose $\rightarrow 1-5$ mg/kg/day

(2) Intracranial hemorrhage

(Only subdural hemorrhage is common in term baby).

- Capillaries in the subependymal germinal matrix is fragile; so they rupture.

Preterm; sudden pale; Shock, fontanel bulging; seizures - Intra ventricular hemorrhage (IVH)



Risk IVH preterm - < 1500 gm $\rightarrow 30\%$

[50% IVH \bar{c} in 24 hrs.

[75% " " 72 hrs.

IOC for newborn having seizure \rightarrow Trans fontanel USG

Term; Breech \rightarrow IVH

Instrumental delivery \rightarrow IVH



Prevention of IVH:

- Antenatal steroids
- Low dose indomethacin ^{to} baby.
- Prevent acidosis, infections in baby.

③ ~~Asphyxia~~ Asphyxia

Preterm

Term



Periventricular
leucomalacia (PVL)

Parasagittal injury



Power: UL > LL

Spastic quadriplegic cerebral
palsy.



Diplegia

(upper limbs strong
LL are weak)

MRI: Coronal Section

- parasagittal injury.

↳ spastic diplegia type
of Cerebral palsy.

• Mentally Retarded

Q. M/c sequel of Periventricular
leucomalacia in preterm

- Spastic diplegia

MRI: PVL (IOC)

- Less white matter
- Shrinkage Ventricle.

- Mentally Retarded.



Status marmoratus: diffuse neuronal loss following asphyxia.

APGAR Score: 1, 5, 10 minutes

- doesn't help in resuscitation.
- 5'; 7'; 10' low → Worse neuronal outcome.

	0	1	2
A = Appearance	Central cyanosis/ Pale	Acrocyanosis	Pink.
P = Pulse (HR)	0	<100	>100
G = Grimace	No	Grimace	Crying
A = Attitude	Extended	Mid	Flexed
R = Respir effort	Apnea	Gasping	Crying.

NORMAL - 7-10

Moderate to severe asphyxia; out of hospital CPR

↓
ischemia reperfusion injury

↓
Free radical damage.

Rx - Therapeutic hypothermia / Selective head cooling:
33.5°C in 6hrs of life; Keep for 72hrs.

↓
preventive

Hypoxic Ischemic Encephalopathy (HIE):
Injury to brain at severe asphyxia.

Signs	STAGE 1	STAGE 2	STAGE 3
• Loss of consciousness	<u>Hyperalert</u>	<u>Lethargic</u>	<u>Stuporous; Coma</u>
• Muscle tone	<u>Normal</u>	<u>Hypotonic</u>	<u>Flaccid</u>
• Posture	<u>Normal</u>	<u>Flexion</u>	<u>Decerebrate</u>
• Tendon reflexes/ clonus	<u>Hyperactive</u>	<u>Hyperactive</u>	<u>Absent</u>
• Myoclonus	<u>+</u>	<u>+</u>	<u>-</u>
• Moro reflex	<u>Strong</u>	<u>Weak</u>	<u>-</u>
• Pupils	<u>Mydriasis</u>	<u>Miosis</u>	<u>Unequal, poor light reflex.</u>
• Seizures	<u>None</u>	<u>Common</u>	<u>Decerebration</u>
• EEG finding	<u>Normal</u>	<u>Low voltage changing to seizure activity.</u>	<u>Burst suppression to isoelectric</u>
• Duration	<u>< 24 hr it progresses; otherwise may remain @</u>	<u>24 hrs - 14 days</u>	<u>Days to weeks</u>
• Outcome	<u>Good.</u>	<u>Variable</u>	<u>Death, Severe deficit</u>

DOC: Seizures in New born: Phenobarbitone.
(Bolus, 20mg/kg)



③ CVS: Hypotension; PDA (Patent ductus arteriosus).

PDA: Preterm \Rightarrow Asphyxia \rightarrow PGs

Infection \Rightarrow Rubella infection \rightarrow Vessel wall defect.

\downarrow
Rx: Surgery

In preterm: Rx: NSAIDs

(Ibuprofen) \blacktriangleright Indomethacin

\downarrow
Less nephrotoxic

If Medical management fails \rightarrow Sx

O/E of PDA:

- 6-10 wks of life CHF
- Preterm baby, failure to wean off ventilator (hypoxia; CO₂ retention)

O/E: • Bounding pulses & wide pulse pressure.

- Continuous machinery murmur at the upper left sternal border.

④ GI System: Necrotising Enterocolitis

⑤ Eye: ROP/Retrolental fibroplasia.

⑥ Hypothermia; ^{Hypo}Glycemia; HypoCa; HypoMg.

⑦ Anemia; jaundice; Infection.



Necrotizing Enterocolitis (NEC):

R/F: ① Immature Gut.

NEC: Susceptibility of premature infants

- Reduced proteolytic enzymes
- ↑ Gastric pH
- ↓ peristalsis
- ↓ motility
- Altered epithelial membr^r & tight junction
- Altered bacterial flora.
- ↓ mucous coat
- Altered mucous protein
- ↑ epithelial permeability.

- Sepsis toxins
- Top fed (Cow milk)

② Mature Cocaine

③ PPIs ; Anti H₂

④ Rapid advancement of feed.

Prevention: ① Antenatal steroids

- ↓ IVH
- ↓ PVL
- ↓ NEC
- ↓ RDS
- ↓ Neonatal mortality.

HUMAN MILK: ↑ proteolytic enzyme

↓ Gastric pH

↑ peristalsis

↑ motility

Less pathogenic bacterial flora



- Altered mucus coat (Improves)
- ↓ epithelial permeability.

② Trophic feeds - expressed milk
10-15 ml/kg/day.

③ Avoid PPIs ; anti H₂

④ Avoid rapidly feed advance.

⑤ Probiotics.

Q. 1 yr old infant; 10-12 episodes of watery stools / day for last 9 days. Along c Zn, which else should be advised -

A) ORS c antibiotics

~~B) ORS orally.~~

C) ORS c low lactose diet

D) ORS c low lactose diets & probiotics.

PROBIOTICS:

Prevents NEC - *Lactobacillus acidophilus*
& *Bifidobacterium infantis*
to VLBWs

- may use in Rotavirus:

Lactobacillus rhamnosus
& *Saccharomyces boulardii*

HMT = Hyponatremia, Metabolic acidosis, Thrombocytopenia.



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Modified Bell's Staging for NEC:

Stage Ia: Suspected NEC	Distention; ileus; occult blood in stool.
Ib: Suspected	Gross blood loss.
IIa: Definite	Focal pneumatosis
IIb: Definite	HMT, Diffuse pneumatosis; Portal Venous Gas.
IIIa: Advanced	DIC; Shock; Peritonitis
IIIb: Advanced-perforation	Pneumoperitoneum

Q. Neonate; distended abdomen & B/L gas shadow under the diaphragm.

Δ - NEC

↳ In 90% preterm

2nd-3rd wk of life.

Q Features of NEC are all except

A) Abd. distention

B) ↑ Bowel sound

C) Pneumoperitoneum

D) Metabolic acidosis

R_x: NEC

- Stop all oral feeds
- TPN (Glucose, AA's, lipids)
- Antibiotics (Cefotaxime; Vancomycin; Metronidazole)
- Stage III - may require Sx.



Q. Child c NEC c perforation & poor general condⁿ is treated c :

- A) Conservative t/t only.
- B) Frank drain c glove
- C) Laprotomy c resection anastomosis
- D) Extracorporeal membr^r oxygenation.

Stage III < Stable - Laprotomy
Unstable - Peritoneal drain

NEONATAL SEPSIS:

= Symptoms + Bacteremia

EARLY

< 72 hrs

LATE

> 72 hrs.

R/F - Maternal Fever < 7 days
Foul liquor
PPROM: Chorioamnionitis.

M/c/c - Nosocomial
M/c/c world - Coagulase -ve staph
In India - Klebsiella, S. aureus.

M/c/c world: Group B streptococcus, E. coli
(M/c)

Meningitis => CSF examⁿ

Rx - Ampicillin + Gentamycin

- Cefotaxime + Amikacin

M/c/c India: Klebsiella, S. aureus

Sepsis screen for early diagnosis:

- ① TLC < 5000/cumm or > 20000
- ② ANC < 1500/cumm
- ③ PS for band cells / Immature neutrophils > 20% (I/T > 0.20)
& toxic granules
- ④ Micro ESR (6mm^N - 3 days of life) > 15mm fall in 1st hour
- ⑤ CRP; Procalcitonin +ve
- ⑥ Lumbar puncture (In late sepsis)



⑦ Chest X-Ray

ANC = Neutrophil + Band cells

Q Lab finding in Neonatal Sepsis except -

- A) ↑ CRP
- B) Leucocytosis
- C) ↓ ESR
- D) Toxic granulated multilobulated nuclei

Duration of Antibiotics in Neonatal Sepsis:

Bacteremia = 10-14 days

Meningitis = 21 days

Arthritis, Osteomyelitis = 4-6 wks

Temp^r regulation of newborn:

Non-shivering Thermogenesis -
Brown Fat

- Nape of Neck
- Interscapular
- Around kidneys & adrenal
- Around blood vessels (around mesentery).

Axillary temp^r:

- Normal 36.5 - 37.5°C
- Cold stress / mild hypothermia (36 - 36.4°C)
- Moderate hypothermia (32 - 35.9°C)
- Severe (< 32°C)
- Hyperthermia > 37.5°C - mostly iatrogenic.

Prevention of hypothermia: KMC (Kangaroo mother care).



Q. All components are of KMC except:

- A) Kangaroo position
- B) Kangaroo nutrition
- C) Early discharge & follow up.
- ~~D) Supplementary nutrition.~~

Q. When to stop KMC:

- When a child reaches 2500gm wt.

Q. Delivery room temp - $> 72^{\circ}\text{F}$ (AAP)

$> 25^{\circ}\text{C}$ (WHO)

$\hookrightarrow (25-28^{\circ}\text{C})$

ALIMS NICU protocols

- $< 28\text{wks} / < 1000\text{gms} \rightarrow$ put baby in Polythene bag.

\downarrow
reduces convection

- Incubator \rightarrow Reduces convection.

Q. Mechanism of heat transfer in overhead radiant warmers :-

Radiation + \downarrow convection loss.

Hypothermia:

CNS: Asphyxia

CVS: Asphyxia Cardiomyopathy

Lungs: ARDS

Kidneys: Asphyxia ATN

Hypoglycemia; Hypo Ca; Hypo Mg.

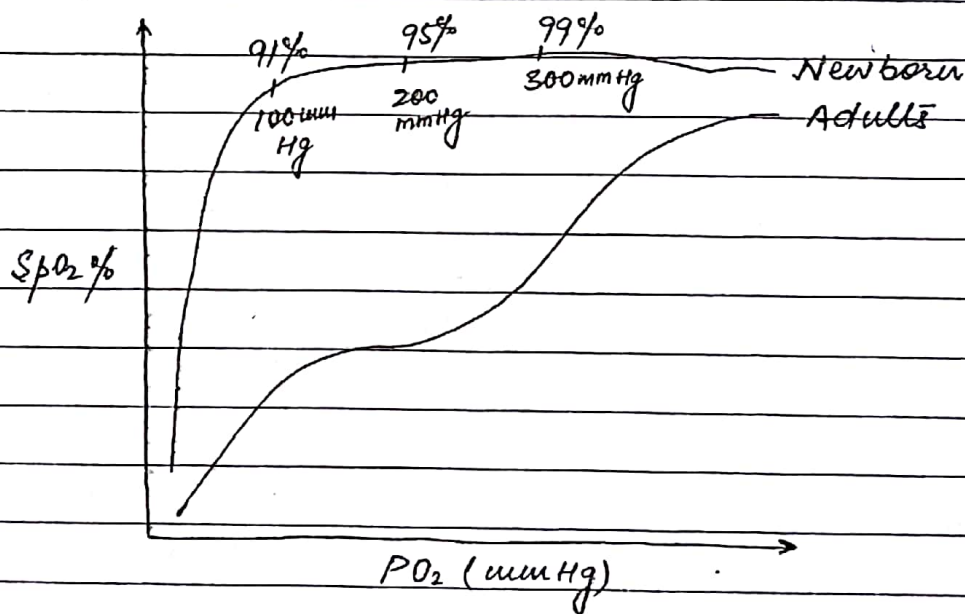


Retrolental Fibroplasia / ROP (Retina of Prematurity):

R/F: Preterm.

High flow O_2 .

- Proliferation, dilatation & tortuous vessels.
- Tractional retinal detachment.
- ROP stage I-V: 'plus' - Blindness.
- Stage IV: Incomplete tractional RD
- Stage V: Complete " "



WHO targets $SpO_2 = 91-95\%$ in preterm.

BLINDNESS: ^{Inv. of ROP} Regular indirect ophthalmoscopy.

AAP ROP guidelines - (Risk < 30 wks / < 1500 gms)

Gestation (wks) 1st visit to ophthalmologist

Post menstrual (wks)

Interval (wks)

22

31

9

23

31

8

24

31

7



25	31	6
26	31	5
27	31	4
28	32	4
29	33	4
30	34	4

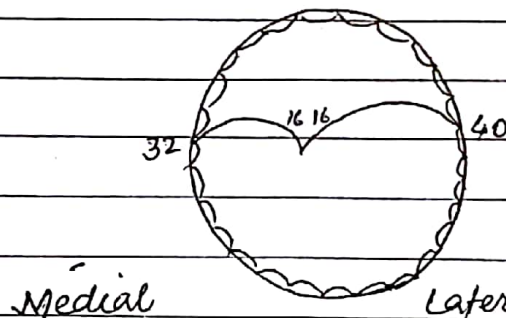
Q. Pediatrician in a district hospital calls ophthalmologist for-

a) New born c Respiratory distress

b) New born 28 wks gestation

↳ After first visit, baby has to go to ophthalmologist, every 2 wks till his/her eye look like term retina.

Retinal Vascularization: Left eye.



Vascularization of Retina
Starts at 16 wks,
finishes medially at
32 wks & laterally
at 40 wks.

Rx: ROP

- Laser Photocoagulation / Peripheral ablation
- Type I ROP (all plus disease)
- Cryotherapy.
- Stage V → Retinal reattachment.

New drug - Bevacizumab → Anti-VEGF; resistant.



#

Respiration :

Tachypnea : RR > 60/min.

Silverman Anderson Retraction Score :

Feature	Score 0	1	2
Chest movement	Equal	Resp ^r lag (upper chest inspiration)	Paradoxical/ Seesaw Resp ^r
Intercoastal Retraction	None	Minimal	Marked
Xiphoid Retraction	None	"	"
Nasal Flaring	None	"	"
Expiratory Grunt.	None	Audible c Stethoscope	Audible.

#

Downes scoring for Respiratory distress :

Feature	Score 0	1	2
RR (per min)	< 60	60-80	> 80/apneic episode
Cyanosis (Central)	None	In room air	in 40% O ₂
Retractions	'	Mild	Moderate - Severe.
Grunting	'	Audible c Stethoscope	Audible c Stethoscope
Air entry (mid-axillary line)	Clear	↓ (Delayed)	Barely audible.



M/c/c of RD \rightarrow Surfactant deficiency

Surfactant \rightarrow Phosphatidyl choline (65%) (Most imp.)

Phosphatidyl glycerol

Phosphatidyl inositol

Phosphatidyl ethanolamine

SP-A, SP-B, SP-C, SP-D.

Other protein

\hookrightarrow Homogenates 20 wks gestation

Amniotic fluid - 28 wk gestation

Mature levels - 95% 35 wks.

RDS risk \propto Degree of prematurity.

Risk of RDS:

< 28 wks - 60-80%

> 37 wks - < 5%

Infant of diabetic mothers ^{at} term

Rare in LUGRs \rightarrow Stress \rightarrow Cortisol

Lecithin: Sphingomyelin ratio

> 2 \rightarrow lungs mature.

> 3.5 infants of DM mothers.

CxR: RDS (lungs): Ground Glass appearance.
(White out lung).

Air bronchogram - Classic feature of RDS.

(N) CxR seen in Early RDS.



Rx: • Humidified O₂ (40-60%)

- We don't give 100% O₂ bcoz of ROP.

• Nasal CPAP: mild - moderate distress
early in ELBW's

FiO₂: Start @ 40-60% ----> room air

PEEP: 5cm H₂O

• Intra-tracheal surfactant - Severe; as rescue.

- Intubate the baby

Give Surfactant

Extubate the baby

IN
SUR
E

Survanta - Bovine

Curosurf - Porcine

Surfasurf - Calf.

Synthetic also available

Q. 32wks, Preterm baby in emergency C.S.

Grunting, RR = 70/min. Best management of choice

A) Humidified O₂ by hood

B) Mechanical ventilation.

C) CPAP

✓ D) Surfactant therapy & mechanical ventilation.

Q. All occur in RDS except:

A) Cyanosis

B) Occurs in preterm

C) More in IDM

✓ D) Treated by 100% O₂



Q. All true about CPAP except:

- A) Initiated FiO_2 0.40 - 0.60
- B) Used in apnea of prematurity.
- C) Improves compliance.
- D) Volume; $FRC \uparrow$ [RDS $FRC < CV$]
- E) Used prophylactically in ELBW's.
- ✓ E) All true.

Q. Term female, Birth wt. = 3.5kg, uncomplicated delivery.

Respiratory distress after birth.

CxR - Ground glass appearance.

On ventilation & given surfactant.

but condⁿ deteriorates & hypoxemia increases.

H/o sibling dying in one week & similar complain. ECG & blood culture - (N).

↳ Δ = Neonatal Pulm. Alveolar proteinosis

↳ Autosomal Recessive

↳ Mutation in Protein B (Rarely in C)

Rx: Early lung transplantation

Postmortem Bx - Pink eosinophilic material in lung.

- Idiopathic: 90%

Adult or acquired

IgG antibodies to GM-CSF

- Secondary - 5-10%

Haematological malignancy

Inhalational lungds.

Silicon

Titanium oxide.

- Congenital: 2%



CT scan - Crazy paving pattern
(Prominent intraalveolar septae).

Rx - Early lung transplant in neonates.
Adults → Broncho alveolar lavage.

Respiratory distress: Newborn

Tracheo-esophageal fistula (TEF)

M/c type - Type C

Distal TEF

Esophageal atresia

- Cyanotic newborn & frothing
- Aspirates gastric juice lead to pneumonia.
- Not a surgical emergency.

Rx - Keep him propped up.
- Suction catheter in upper blind esophageal pouch.



Diaphragmatic hernia:

85% cases - left.

pushed heart & trachea - to opposite side.

- Scaphoid Abdomen
- Barrel chest
- Mediastinal shift to right.
- Apparent dextrocardia
- Peristalsis on left chest.

Diaphragm - develops from septum transversum
& pleuroperitoneal canals.

↳ Fail to close on left side
(Bochdalek hernia)

- < 5% are R/L



Q. Cause of death in Congenital Diaphragmatic Hernia:

- A) Septicemia
- B) Pulmonary hypoplasia - Left
- C) Hemorrhage.

Bag & mask is absolutely c/i bcoz it will cause further abdominal distension.



So elective intubation is done.

Baby born → Bag & mask c 100% O₂



diaphragmatic hernia



Intubate

Next step - NG tube to decompress the gut.

Born baby → Diaphragmatic hernia



Intubated



Heart further to right

Next step - Remove the tube
& Reintubate.

Q. Most imp. Prognostic factor in Congenital diaphragmatic hernia:

- A) Pulm. HTN - Persistent (PPHN)
- B) Age
- C) Time of Sx
- D) Size of defect.



Prevention : PPHN

- Elective intubation
- HFOV > 300-600/min

Rx : PPHN

- iNO \rightarrow Pulm. vasodilator.
- Sildenafil PDE-V inhibitor

New drug : Bosentan, Ambrisentan — Endothelin antagonist.

- $PGI_2 \Rightarrow$ Iloprost
- Amlodipine ; ECMO (Extracorporeal memb^r oxygenation)

Transient Tachypnea of Newborn (TTNB):

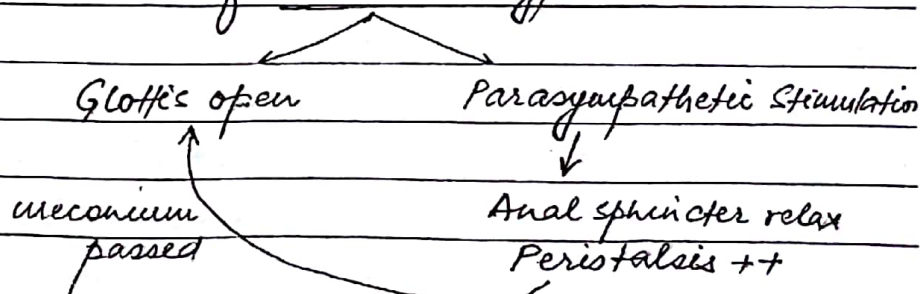
- R/F :
- Term by C.S. (lungs are wet)[±]
 - Macrosomia[±]
 - Excessive maternal sedation.
 - Precipitous labour.

CxR — a prominent horizontal fissure
 \hookrightarrow Most specific feature.

- Benign condⁿ
- Self limited : 48-72hrs.
- FiO_2 requirement < 0.40
- Never require mechanical ventilator.

Meconium Stained liquor :

Meconium is marker of Perinatal Hypoxia

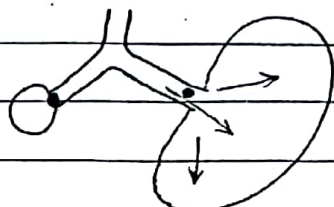




Perinatal hypoxia: Common in posterior
6coz of UPL.

Meconium:

Physical -



- ball valve mechanism

- air leak 20-30%

Chemical - Irritant \rightarrow pneumonia
 \hookrightarrow Impair surfactant funcⁿ.

Biological - Good culture media

Meconium Stained liquor.

Baby born

Vigorous

Tone is good

Respr effort is good

HR > 100 /min

yes

No

Transfer
the baby to
mother

Catheter
in the nose

PPV @ 100% O₂.

Q Sequence of Resuscitation

A) Mouth \rightarrow Nose

B) Nose \rightarrow Mouth

C) Mouth \rightarrow Nose \rightarrow Trachea



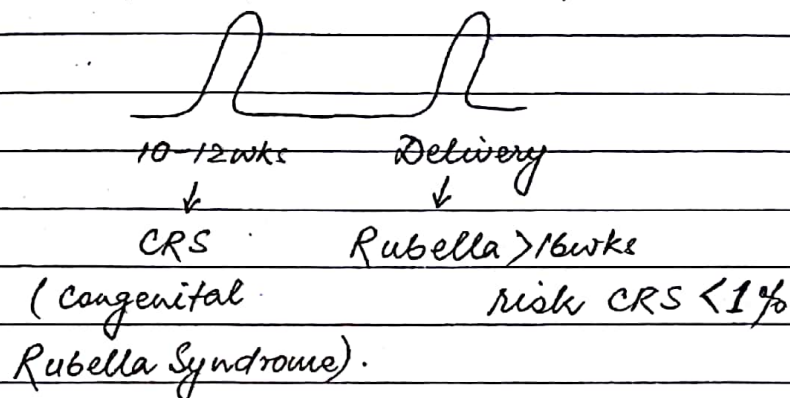
Intra uterine TORCH infections:

Others - HIV, HBV, Varicella, Syphilis.

Common features:

① - Asymptomatic

Rubella: Transmission - 2 peaks



② - If Symptomatic

- SGA; failure to thrive
- Anemia; thrombocytopenia
- Hepatosplenomegaly
- Unexplained Rash & cholestasis.

Δ - IgM (Active of infection)

IgG persisting beyond 6-9 months

HIV in infants

- Infants can't make IgM-HIV or IgA-HIV.
- Maternal IgG-HIV can persist in the baby for 18 months.
- HIV < 18 month: Diagnosis.

Best: DNA & PCR

P24 assay

Culture is difficult.



Intracranial calcification & Chorioretinitis] M/c/c - Toxoplasmosis
- CMV

Toxoplasmosis

- 25-50%
- Choroid plexus calcification.
- Subependyma & Caudate nucleus calcification.
- ~~Hydro~~ Hydrocephalus
- Seizures.

CMV

- Periventricular Calcification.
- Atrophy of brain. (Cortical).
- Hydrocephalus ex vacuo.
- M/c/c of Non syndromic SNHL
- Seizures
- Microcephaly
- Mental Retardation.

Q. Pregnant lady; no complain
Mild cervical lymphadenopathy
in 1st trimester. Prescribe

prevent Vertical Transmission \rightarrow Spiramycin but she was non compliant. Baby born

hydrocephalus &
intracerebral calcification.
 \rightarrow Toxoplasmosis

DOC: Rx: Pyrimethamine
+ Sulfadiazine.

Δ : Best: IgM Immunosorbent
assay.

Sensitivity of ELISA:
IgA \gg IgM

Q. True about transplacental
CMV infection:

- It is M/c/c of non-syndromic SNHL.

Q. Doesn't establish Δ of CMV
in neonate -

- A) Urine culture of CMV
- B) IgG CMV antibodies in blood
- C) Intra-nuclear inclusion bodies in hepatocytes (Owl-eye)
- D) CMV viral DNA in blood by polymerase chain reacⁿ.

Best specimen - Urine culture & Saliva.

CMV disease: Retinitis, colitis, pneumonia.



Rx: CMV

DOC: i.v. Ganciclovir (Severe, child, pregnant)

- Oral; Prodrug → Valganciclovir
- Resistant to oral Foscarnet.

Congenital Syphilis:

EARLY

- C in 1st 2yrs of life.
- Mucocutaneous rash/rhinitis (Snuffles).
- Lymphadenopathy.
- Hematological (Autoimmune Anemia)
- Renal lesion
- Skeletal (Osteochondritis, metaphysitis, Periostitis)
- Glaucoma
- Pseudoparalysis of Parrot.

LATE

- After 1st 2yrs.
- Hutchinson's Triad:
 - ① Hutchinson teeth/
mulberry molars - 1st
lower molars.
 - Saddle nose, Frontal bossing,
Olympian's brow,
 - Hogoumeraki's Sign
(sternoclavicle prominence)
 - Rhagades
- ② Interstitial Keratitis
- ③ Nerve deafness (SNHL)
- Clutton's joints
(painless joints)
↳ Risk of injury.

Pseudoparalysis:

M/c/c - Scurvy

- Early syphilis
- Osteomyelitis
- Septic arthritis.
- Hypokalemia → Hypotonia.

DOC - Penicillin G
→ 10-14 days



Rubella Syndrome:

Ina'id - Microcephaly (Mental Retardness)

PDA

Cataracts.

M/c eye manifestation of Rubella - Salt & pepper fundus

Cataract

Glaucoma

Micro-ophthalmia

HEART - PDA

Peripheral pulm. stenosis

VSD

ASD (Rare).

Q. Rubella embryopathy except.

A) Deafness

B) MR

~~C) AS~~

D) PDA

Q. True about Rubella embryopathy except:

A) Diagnosed when IgM antibodies in child.

☒ B) Infection after 16 wks results in major congenital anomalies.

C) Deafness, heart disease, Cataract.

Q. Hypoplastic limb - Varicella (Chicken pox) embryopathy during pregnancy.



Varicella Embryopathy -

- Skin Rash
- Optic nerve hypoplasia
- Brain - Cortical atrophy.
- LS plexus - Aplasia / hypoplasia limbs.

Mother get chick pox - 5 days before delivery
or, \bar{c} in 2 days of delivery.



Baby chicken pox illness.



Prevention: Varicella Zoster Ig to the baby.



even after 120 hrs of exposure.

Q Pregnant; HBsAg +ve: no jaundice
HBeAg +ve \Rightarrow 90% chance baby is carrier
& later in life Portal hypertension.
↓
Ascites, Splenomegaly, Varices

HBeAg -ve \Rightarrow Anti HBeAg +ve
then 10% chance of Vertical transmission

Give HB Ig baby to 12 hrs of life
HBV vaccine to baby \bar{c} in 24 hrs of life.

Pricked by HBsAg +ve patient
Are you immunized?
~~Anti HBsAg~~ • Anti HBs Ab titre
Good >10 IU/cul; High risk >100



Incomplete vaccination / Not know titre



HBV + HB Ig
a/c to CDC guidelines.

If titre is good - don't do anything.

#

'CDC' HBV DNA load > 1000 IU/ml

↳ Can't join Surgical branch.

Neonatal jaundice

Hem.



Hemoxygenase
↓ CO₂

Biliverdin

↓ B. reductase

Bilirubin

Et Co. - hemolysis.



Metalloporphyrin

1gm of Hb = 34mg of bilirubin

1gm/dl Albumin binds to 8mg bilirubin

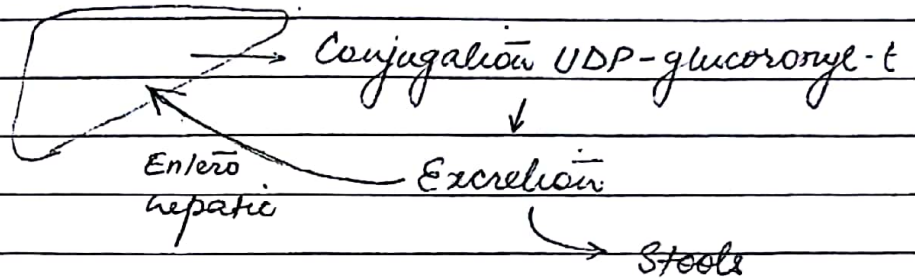
- Unconjugate bilirubin passes BBB & cause jaundice & Kernicterus.
- In 1st 2wks, BBB is not developed properly.
- (N) S. Albumin = 3.5-5.5g/dl
- Healthy term baby can bind 24-25mg/dl bilirubin.
- Sick, preterm, risk factors can go into early Kernicterus.
- In adults ammonia cross BBB in hepatic encephalopathy.
- Healthy Baby (< 1000 g) → We start phototherapy (5-7mg/dl) Bilirubin
- ↓
- Sick baby → Bilirubin (4-6mg/dl) - we start phototherapy.



Bilirubin



uptake by liver (γ -ligand uptake)



$> 2 - 2.5 \text{ mg/dl}$ in adults \Rightarrow Yellow Sclera.

Kramer's zone:

progression of jaundice in new born is
cephalo-caudal
(Bilirubin)

Zone I (5 mg/dl) \rightarrow

Zone II (10 mg/dl) \rightarrow

Zone III (12 mg/dl) \rightarrow

Zone IV (15 mg/dl) \rightarrow

Zone V ($> 15 \text{ mg/dl}$) \rightarrow

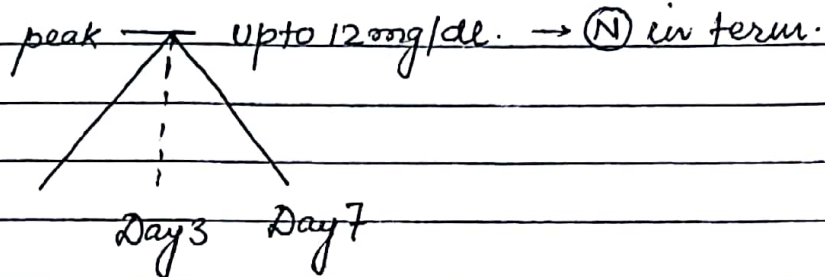
\hookrightarrow Danger Zone.

6 causes of physiological jaundice:

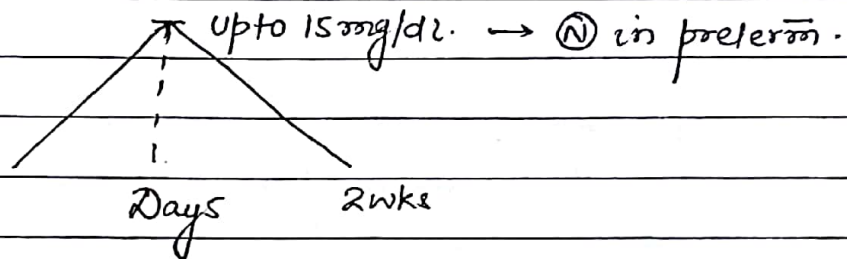
- ① Life span of RBC is less.
- ② Hematocrit more
- ③ Newborn deficient in γ -ligand.
- ④ UDPglucuronyl-t deficient in newborn.
- ⑤ Excretion reduced.
- ⑥ Enterohepatic circulation \uparrow .



Physiological jaundice:
jaundice in term babies.



Preterm — have more jaundice.



Pathological jaundice:
— Hemolysis.

- M/c/c — Incompatibility Rh/ABO [O mother; baby A/B]
- RBC ~~membrane~~ memb^r defect.
 - RBC enzyme defect.

Def: jaundice \bar{c} in 24 hrs of life.

* Rh-ve mother; previous abortions
we take cord blood samples.

- Rh status of baby. → \bar{c} /+ve
- Hb → 10 mg/dl
- Bilirubin → 5 mg/dl.
- Peripheral Smear (P/s) +ve
- Direct Coombs test (DCT) → +ve

↓
means Severe hemolysis



$R_x \rightarrow$ Exchange transfusion at birth.

① jaundice \bar{c} in 24 hrs of life.

② Reaching 20mg/dl

③ Rate rise $>0.2\text{mg/dl/hr.}$

④ Persisting beyond

- 1 wk terms

- 2-3 wks preterm.

⑤ Clay stools.

Photo/therapy:

- Any jaundice on day 1 of life start phototherapy.
Serum bilirubin cut off.

	Photo/therapy		Exchange transfusion	
	Healthy babies	Babies \bar{c} R/F	Healthy babies	Babies \bar{c} R/F
Day 1	\leftarrow Any visible jaundice \rightarrow		260(15)	220(10)
Day 2	260(15)	170(10)	425(25)	260(15)
Day 3	310(18)	250(15)	425(25)	340(20)

R/F ① Gestation <35 wks / wt. $<2\text{kg.}$

② Sepsis

③ Hemolysis

④ Asphyxia

⑤ Sick baby.

Principle:

- Structural isomerization

• Bilirubin \rightarrow LUMIRUBIN

- Photo isomerization

• $4Z15Z \leftrightarrow 4Z15E$ (Soluble).

- Minor pathway \rightarrow photo-oxidation.

40 cm away, falls @ $4-6\text{mg/dl/day.}$



- Phototherapy occurs at 425-475 nm of blue green light.
- Irradiance \rightarrow 6 micro Watt/cm²/nm
- Intensive $>$ 30 micro watt/cm²/nm.

Q

AIIMS Nov. 2013

Which does not effect the efficacy of phototherapy?

- A) Types of phototherapy lamp.
- B) Skin pigmentation.
- C) Spectral radiance of incident light
- D) Initial bilirubin levels.

#

Complication:

- Hyperthermia; insensible losses
- Hypocalcemia
- Diarrhoea.
- Cover eyes & genitals
 - Retinal damage
 - Mutations.
- Phototherapy is C/I in conjugated jaundice
 - \downarrow
 - Bronze baby Syndrome.
(Skin, Urine).

Exchange Transfusion:

- Double volume exchange transfusion.
- # Blood vol. of new born = 80 ml/kg
- $\rightarrow 2 \times 80 \text{ ml/kg} = 160 \text{ ml/kg}$
- Transfuse fresh (< 7 days) whole blood.
 - Reduces bilirubin by 85%
 - \downarrow
 - Not 100% because bilirubin is also in tissue.



- Albumin ↑ ~~efficiency~~ efficiency.

Complications:

- Infections
- ACD (Acid citrate dextrose)



Bicarbonates

1 mole of citrate → 3 mole of Bicarbonate.

- Metabolic alkalosis
- Hypokalemia, hypocalcemia.
- Old blood → Hyperkalemia, metabolic acidosis.

Persistent jaundice:

Cause — Hypothyroidism

Breast milk jaundice

Hematoma (Cephalhematoma, IVH)

Clay colour stool (Cholestasis)

Criegler-Najjar Syndrome type II (Milder form)
↳ deficiency of UDP-glucuronyl-t

Criegler-Najjar Syndrome type 1 — Very Severe
↳ Absent UDP-glucuronyl-t.

Pathological jaundice

Breast milk jaundice

Breast feeding jaundice.

- Onset → day 14

- Onset → Day 3.

- Some mother have pigment

- In primigravida.

& inhibit conjugation.

- Starvation stimulates entero-

- Day 14 = 20-30 mg/dl

hepatic circulation.

- May Kernicterus.

- Persists: 4-6 wks.



Rx: Temporary interrupt
48-72 hrs.

Rx: Ensure feeding.

Meanwhile give
formula milk.

Q

True about jaundice in newborn (neonates) is -

- A) Can be seen after Ventouse delivery.
- B) Physiological jaundice seen \bar{c} in 48 hrs of birth.
- C) Increased conjugated bilirubin leads to kernicterus.
- D) Breast milk jaundice is max^m in 7 days of birth.

Neonatal cholestasis:

Neonate, jaundice & Clay stools.

- Direct bilirubin $> 2 \text{ mg/dl}$ or $> 20\%$ total bilirubin

Medical - Common.

Neonatal hepatitis \rightarrow CMV

Sepsis

Galactosemia

α -1 antitrypsin deficiency.

Neonatal hemochromatosis.

Surgical - Extra hepatic

biliary atresia (EHBA)

Rx - Kasai's surgery \bar{c} in

8 wks of life otherwise

80% die.



M/c/c of indication of
liver transplant in babies

\rightarrow EHBA.

Q

Which is an ominous sign in a 10 day old newborn?

- A) Unconjugated hyperbilirubinemia
- B) Conjugated "
- C) Failure to gain wt.
- D) Doll's eye reflex

\hookrightarrow Normal in 1st 10 days of life.

GGT → Gamma Glutamyl Transferase.



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Medical - Common

Surgical - EHBA

- GGT ten times higher in Surgical causes.
- DO USG → Shows Intra hepatic biliary radicle (IHR) are dilated.
- Triangular cord Sign^g

- HIDA nuclear Scan:

HIDA dye not seen in gut, even in delayed images; white in hepatitis excretion of dye.

Best test - Liver biopsy.



Shows dilatation & proliferation of intra hepatic bile duct.

Before doing HIDA Scan we have to give phenobarbitone 2 to 3 days before test.

Gold Standard - Pre operative Cholangiography.

Alagille Syndrome -

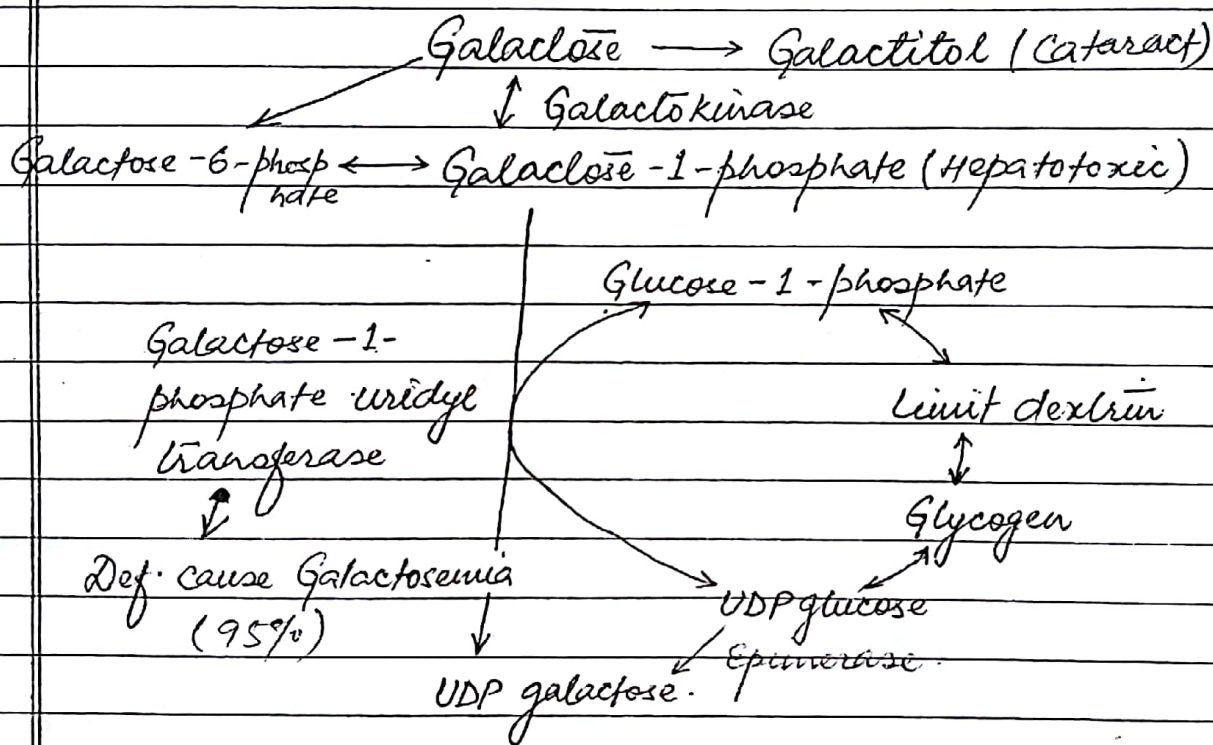
- Cholestasis
- ~~AD~~ AD
- Bile duct paucity Syndrome.
- M/c heart disease → Peripheral pulm. stenosis.

↑
Rubella



- Q. 1 month old child present c conjugated bilirubinemia & intrahepatic cholestasis. On liver biopsy staining c PAS, red coloured granules were seen inside the hepatocytes. Probable diagnosis is -
- α -1-antitrypsin deficiency.
 - Congenital hepatic fibrosis
 - Hemochromatosis
 - Wilson's ds.

CMV - Intracellular "owl eye" inclusion bodies.



- He get jaundice, bleeding, hypoglycaemia, PTM.
- Galactitol cause cataract.
- Rx - Lactose free milk.
- Galactokinase deficiency → Only cataract
No liver failure
- Fructokinase def. → Causes Benign Fructosuria
↳ No symptom.

Fructose-1-phosphate leads to liver failure.



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Q. M/c/c of Neonatal Cholestasis:

- A) EHBA
- B) Neonatal hepatitis
- C) Choledochal cyst.
- D) Physiological

Q. Neonatal cholestasis seen in -

- A) Chronic hepatitis (>6 months)
- B) Hep. B
- C) Galactosemia
- D) Rh incompatibility (cause ~~in~~ unconjugated jaundice)

Q. Pregnant lady, HBsAg +ve, No jaundice.

↓
next step → HBeAg

• HBeAg +ve (90%)
Carrier HBsAg → Portal HTN

↓
Triad - Ascites
 Splenomegaly
 Varices.

• HBeAg -ve
 anti HBe Ab (+)
 ↳ 10%

Immunization [HB Ig - baby c in 12hrs of life
 ↓ [HBV - baby in 24hrs of life
 prevent Vertical transmission



CDC guidelines:

Pricked by HBs Ag ⊕ patient



Are you immunized?



Anti HBs AB titre

Good > 10 mIU/ml

High risk > 100 mIU/ml.

Don't know titre → Incomplete



HBV + HB Ig

If Good titre → Nothing is to be done

Q.



New born, Respiratory distress:

Neonatal Seizures:



Neonatal hypoglycemia:

Causes:

- Limited stores → Preterm, IUGRs
- Stress → Sepsis
- Polycythemia
- Galactosemia - liver failure.
- Low cortisol - CAH (Congenital Adrenal Hyperplasia)
- Hypopituitarism:
 - Low ACTH; Low LH/FSH → Micropenis.
- Hyperinsulinism - In infants of diabetic mother.
 - ↳ Foetus of β -cell hyperplasia.

↓
Fetus Blood glucose ↓
↑

Pederson's hypothesis

- Insulinoma; Nesidioblastosis
- Beckwith Weidman syndrome.
 - ↳ Hemihypertrophy of limb
 - ↳ Macroglossia
 - ↳ Risk of Wilms
 - ↳ Hyperinsulinism



- Q. A term baby to a diabetic mother, few hours after birth was lethargic & his blood glucose was 30 mg/dl . What should be done next -
- A) Give 10% dextrose orally.
 - B) 10% dextrose i.v. - Bolus 2 ml/kg → Glucose drip
GIR
↓
 $6-8 \text{ mg/kg/min}$
 - C) Give expressed breast milk
 - D) DO exchange transfusion.

Neonatal hypoglycemia:

- | Symptomatic | Asymptomatic |
|--|--|
| → Bolus 2 ml/kg 10% dextrose | → Blood glucose $< 20 \text{ mg/kg}$ |
| fb by Glucose drip | ↓ |
| $6-8 \text{ mg/kg/min}$ | Glucose drip ($6-8 \text{ mg/kg/min}$) |
| | → Blood glucose = $20-45 \text{ mg/dl}$ |
| | ↓ |
| | Breast feed; 1hrly. |

- # A baby of glucose drip & he gets seizures
how to ↑ glucose rate
- Upto 12.5% through peripheral vein.

Max^m glucose infusion rate (GIR) = 12 mg/kg/min .

Emergency drug = i.m. Glucagon.
↓
for hypoglycemia

Glycogenolysis
+ Gluconeogenesis.



Q. Gluagon is effective for Mx in persistent hypoglycemia in all except:

- A) large date for baby.
- B) Nesidioblastosis
- C) Galactosemia
- D) Infant of diabetic mother.

Q. 1 yr old, hypoglycemia & hepatomegaly
No jaundice.

Hypoglycemia doesn't respond to glucagon.



Δ → Von Gierke's (Glycogen storage ds - I)

Glycogen ——— Glucose - 6-PO₄

Von Gierke's $\xrightarrow[\downarrow]{\ominus}$ Glucose - 6-phosphatase.
Glucose

Types of Glycogen storage disorder:

V = Von Gierke's → Liver primary

P = Pompe's ds → Heart primary; Cardiomegaly.

C = Cori → Debranching enzyme. Large QRS complexes.

A = Anderson → Branching " deficiency

M = McArdle's → Muscle phosphorylase deficiency.

Harding =

Tom =

jaundice never occurs in Glycogen storage ds.

R. pompe's: Enzyme replacement therapy.

Enzyme absent in pompe's → lysosomal α-1,4-glucosidase
— Also called Acid & neutral maltase.



Muscle affected in GSD:

Calcium:

(N) S. $\text{Ca}^{2+} \rightarrow 9-11 \text{ mg/dl}$.

Neonatal hypocalcemia:

- S. $\text{Ca}^{2+} < 7 \text{ mg/dl}$
- ~~Best~~ Best index of body calcium

↓
Ionized $< 4 \text{ mg/dl}$ (or) $< 1 \text{ mmol/L}$

- Tetany is rare in infants.
- Tremors, seizures, jitteriness.
 - ↳ Tremulousness & is stimulus sensitive.
 - ↳ Can stop on passive restrain.

Early Hypo Ca^{2+}

Causes - Prematurity
Asphyxia

↳ Infant of DM mother.
Test:

Blood glucose
+ S. Ca^{2+} & S. Mg^{2+}

Late Hypo Ca^{2+}

Cause - Feeding $\bar{\text{c}}$ phosphate rich milk (Cow milk)

Good Ca^{2+} Supplement ($\text{Ca}^{2+}/\text{P} > 2$)

100 ml	Cow milk	Breast milk
Ca	118mg	34mg
PO_4	100mg	15mg



Advantages of Breast milk:

- protects against late onset hypocalcaemia.
- protect against pneumonia

NEC (Necrotizing Enterocolitis).

Allergy, Eczema, asthma.

Rota virus diarrhoea.

Bronchiolitis (IgA-RSV)

Q. Milk deficient in:

a) Iron & Vit. C

Q. Breast milk has enough iron & Vit. C for 6 months.

↓

∴ Scurvy never occurs in ^{1st} 6 months of life.

Q. If baby on exclusive breastfeed for 2 yrs.

↓

Iron deficiency anaemia.

Q. APP recommends Vit. D to all infants - Vit. D drops.

- RDA vit. D infant 400 IU/day

- Breast milk 25 IU/L.

M/c/c of HypoCa^{2+} in infants = Maternal deficiency of Vit. D.

25 [OH] Vit. D = Status.



Infant of DM mother:

- Can be stillborn; preterm
- Macrosomia
 - ↳ Bcoz of Hyperinsulinism
- Linear growth in utero depend upon insulin & or Insulin like growth factor.
- LUGR → White's classification class F/R mother → Placental Vasculopathy.
- Hypoglycemia
- Hypo Ca^{2+} , Hypo Mg^{2+}
- Neonatal jaundice
- Polycythemia → Renal Vein thrombosis (RVT)

Q.

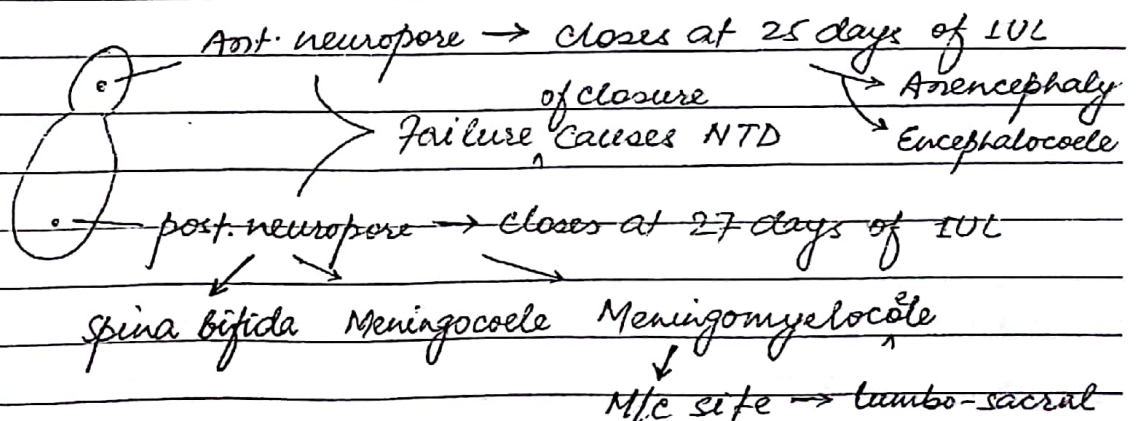
Att. Not seen in infant of diabetic mother:

↓
Hyperglycemia.

Anomaly:

- (Mk) - CVS - 8.5% [VSD; HOCM] Asymmetrical septal hypertrophy
- Neural tube defect → 5%
 - Lazy left colon syndrome → Pseudoobstruction of colon
 - Sacral agenesis / Caudal regression Syndrome
 - ↳ Most specific.

NEURAL TUBE DEFECT (NTD)





Anencephaly:

- Ant. neuropore fails to close.
- No brain, absent of part of hind brain.
- Earliest abnormality diagnosed by USG (10-12 wks) of gestation.

- Most severe NTD
- Don't resuscitate
- Mostly post-term.

Herniation of brain tissue - Encephalocele.

Lumbo-sacral myelomeningocele:

Complication:

- Paraplegia/Paraparesis
- Neurogenic bladder → CKD.
- Constipation (Severe)
- Associated hydrocephalus. (Obstructed)



Because of Arnold-Chiari type II malformation



MRI Brain

Ruptured myelomeningocele:

Best test - Blood culture.

- Cover with Normal saline soaked gauze.
- 95% of neonatal meningitis have leukemia.
- M/c/c of meningocele - Folic Acid deficiency.



Folic acid deficiency:

- -1/+3 months conception - start folic acid.
- 400 mcg given.

Recurrence:

- 1 child - 3.5% chance
- 2 child - 10%
- 3 child - 25%

To prevent recurrence → Folic acid 4mg
↓
reduces risk by 75%.

Δ: ① USG

② Amniocentesis

↳ Acetylcholinesterase

& α-fetoprotein are markers.

α-fetoprotein in mother serum is marker of Neural tube defect.

Resuscitation:

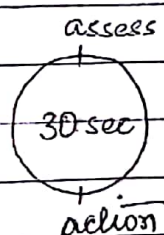
NRP Guidelines 2015:

T = Temp^r

A = Airway: position neck, suction

B = Breathing

C





Indication of Bag & mask \bar{c} 100% O_2 :

- Apnea/gasping ~~after~~ after initial steps.
- HR $< 100/\text{min}$ after 30sec PPV.
- Central cyanosis despite 100% O_2 .

- Chest compression if HR $< 60/\text{min}$, falling after 30sec PPV.
- Chest compression : Bag mask = 3:1

In 1 minute = 120 events

90 Chest compression & 30 bag & mask.

Compression to Ventilation ratio:

- Children/Infants — Single rescuer 30:2
2 rescuer 15:2

- Adults — 1 or 2 rescuer 30:2

CPR sequence \rightarrow CAB

Drugs for resuscitation:

- 1) 0.9% NaCl 20ml/kg bolus \rightarrow Shock
- 2) 1:10,000 epinephrine 0.1-0.2ml/kg



If HR = 0 or falling.

- 3) I.V. $NaHCO_3$ — documented metabolic acidosis.
- 4) I.V. Naloxone — mother opioid addict.

Targeted preductal SpO_2 after birth:

1 min = 60% - 65%

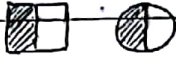
2 " = 65 - 70 %


3 " = 70 - 75 %





4 min = 75-80%
5 min = 80-85%
10 min = 85-90%


GENETICS


 → Heterozygotes for AR carrier


 → Carrier for sex-linked recessive


 → Death


 → Abortion of stillbirth
Sex unspecified.


 → Proband.


 → Consanguineous marriage

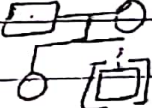
 → Dizygotic twin


 → Monozygotic twin

 → azoospermia

 → Endometriosis

 → Infertility.

 → Adopted in

 → Adopted in

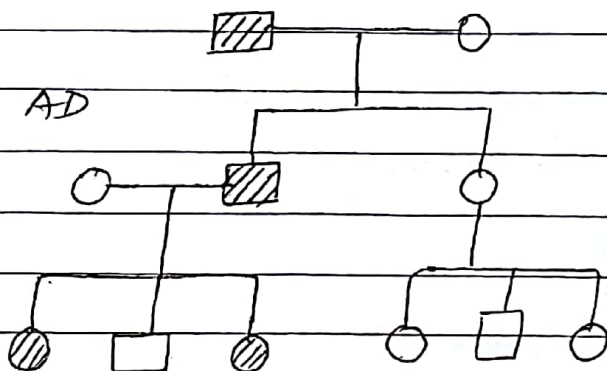


□ —|— ○ → No children for choice
or reason unknown

□ → Vasectomy

○ → Tubal

□ —/— ○ → Divorce.



Autosomal dominant:

D = Dystrophy myotonic → distal myopathy.

O = Osteogenesis imperfecta

M = Marfan Syndrome

I = Intermittent porphyria

N = Noonan Syndrome → Turner phenotype XX & XY.

A = Adult PKD, Achondroplasia

NF = NF (Neurofibromatosis)

T = Tuberculous sclerosis

VH3 = Von Willibrand Syndrome

Huntington's chorea

Familial Hypercholesterolemia

Hereditary spherocytosis.



Turner Syndrome

- 60% case - XO
- Webbed neck
- Cystic hygroma
- Lymphedema of hand & feet.
- Primary amenorrhoea during puberty.
- Streak ovaries
- Cubitus valgus.
- MR rare.
- M/c heart ds - Bicuspid aortic valve & aortic stenosis
↳ Half to one-third.
- 20% coarctation.
- Girls infertile

Noonan Syndrome.

AD

 $XX = XY$

- 25% MR
- Valvular pulm. Stenosis.
- HOCM
- ASD.
- Girls are mostly fertile.
- Boys - Cryptorchidism.
- Clotting factor deficiency.

Q.

Chance of child being not affected if both parents are affected & Achondroplasia is -

A) 0%

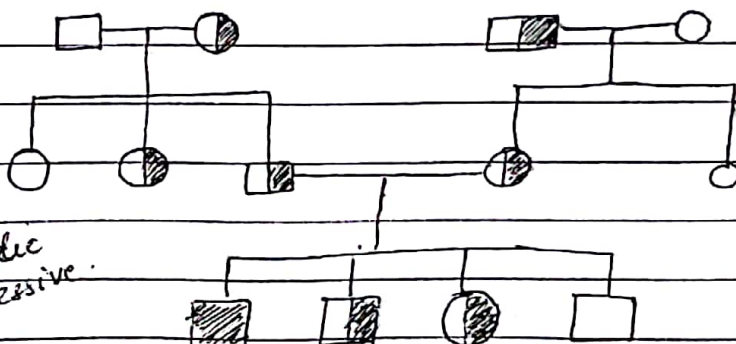
✓ B) 25%

C) 50%

D) 100%

 $AA^c \times AA^c$ (AA) $AA^c \quad A^cA \quad A^cA^c$

Autosomal
Recessive
metabolic
recessive.





Autosomal Recessive:

- Cystic Fibrosis
- α₁ AT deficiency
- Wilson's ds
- Haemochromatosis
- Friedrich's ataxia
- Gaucher's ds
- Niemann's pick ds.
- Tay Sachs ds.
- Hurler's Syndrome.

Mucopolysaccharoidosis (MPS)

- Child c noisy breathing
- Coarse facies.
- Abn accumulation of glucose aminoglycans.
- Gargolism
↳ Chronic rhinitis.

Mucopolysaccharoidosis type I → K/A HURLER'S Syndrome

↓
deficiency of L-Iduronidase.
Rx - Enzyme replacement therapy.

HURLER

- MPS - I
- AR
- Corneal Clouding

HUNTER Syndrome.

- MPS - II
- XL recessive
- Cornea clear.



Enzyme Replacement therapy:

1st to be treat → ① Gaucher's ds

(β glucocerebrosidase)



Company name — GENZYME

CER-zyme.

② Pompe's

③ Hurler (MPS-I)

④ MPS-VI (Maroteaux-Lamy)

⑤ X-linked recessive Fabry's
(CNS & kidney problem)



Lysosomal disorder.

Cherry red spot macula seen.

Gene therapy:

1st to be treated by Gene therapy → X-linked recessive

Severe combined immunodef. (SCID)

(Adenosine deaminase deficiency)

#

British guy

British lady

Chance of child having cystic fibrosis

UK/Europe/ → $\frac{1}{25}$ carrier cystic fibrosis.

askenazi jews

$$\frac{1}{25} \times \frac{1}{25} \times \frac{1}{4} = \frac{1}{2500}$$

AAC × AAC

(AAC) · ACA AAC AA

$\frac{1}{4}$

#

British guy

British lady

(brother died of cystic fibrosis)

Chance of child to have cystic fibrosis

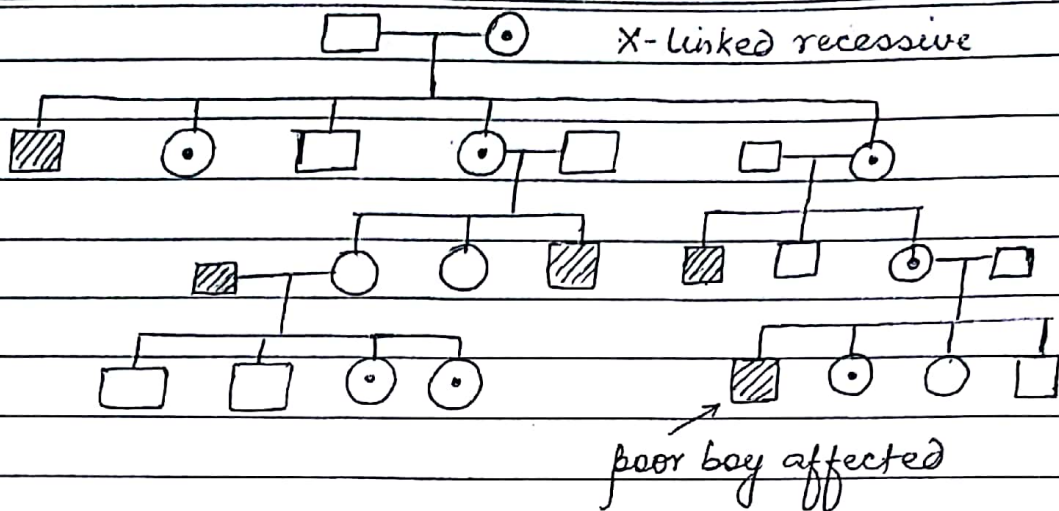
$$\frac{1}{25} \times \frac{2}{3} \times \frac{1}{4} = \frac{1}{150}$$

AAC × AAC

(AAC) · ACA AAC AA



Date / /
Page



$X^c X^c$	$X^c Y$
XY	XY

X-linked Recessive: (Poor ~~lunch~~ boys)

- Duchenne muscular atrophy. (M/c hereditary Neuromuscular)
- Hemophilia A & B.
- G-6PD deficiency.
- Wiskott-Aldrich syndrome.
- Colour blindness.
- Lesch-Nyhan syndrome.
- Chronic granulomatous disease.

Duchenne muscular hypertrophy (DMH):

- Pseudohypertrophy of calf muscle
bcz of fat deposition.
 - Proximal muscle weakness
 - Gower's Sign \rightarrow Not specific.
- $\Delta \rightarrow$ CPK = 10,000 IU

Valley Sign \Rightarrow Hypertrophy of Supraspinatus
Atrophy of Infraspinatus

Also seen in DMH.

↳ specific for boys who have calf hypertrophy.



Human Genome:

- 30,000 gene.

Largest gene - Dystrophin

- ↳ Skeletal muscle
- ↳ Heart - Cardiomyopathy.
- ↳ Brain → $\frac{1}{3}$ cases MR.

Boys - Duchenne - die teens due to recurrent chest infection.

Duchenne → XLR, $\frac{1}{3}$ de novo mutation.

Becker's dystrophy:

- Similar to duchenne
- Mild form & present late.
- X-linked recessive.

Wiskott-Aldrich:

- X-linked recessive
- Eczema
- Thrombocytopenia
- Immunodeficiency.

Chronic granulomatous di-

- X-linked recessive
- Immunodeficiency.
- ~~Also~~ NADPH oxidase deficiency
- Dx → NBT dye test

Lesch-Nyhan Syndrome:

- X-linked ~~recessive~~ recessive
- Purine defect
- HGPRTase deficiency.
- Hyperuricaemia > 6.5 mg/dl
- Self-mutilation → nose, palate, fingers.

✓ A) Serum uric acid. \xrightarrow{H} Lesch Nyhan Syndrome.

C) S. LDH

A) Hypercalcaemia \rightarrow 24hr urine $\text{Ca}^{2+} > 4\text{mg/kg}$

✓ Similar presentation in father

Urolithiasis

Fanconi Syndrome - Proximal tubular defect

60% >85% >88% 100%

↳ Oculo-cerebro renal gene. (OCTL gene).

Microcephaly
MR

Fauci's Syndrome.

Fructose-1-phosphate
Glucose-1-phosphate } proximal tubular toxic.



Fanconi's Syndrome:

- Genetic

↳ X-linked recessive - Lowe

XLR Dent

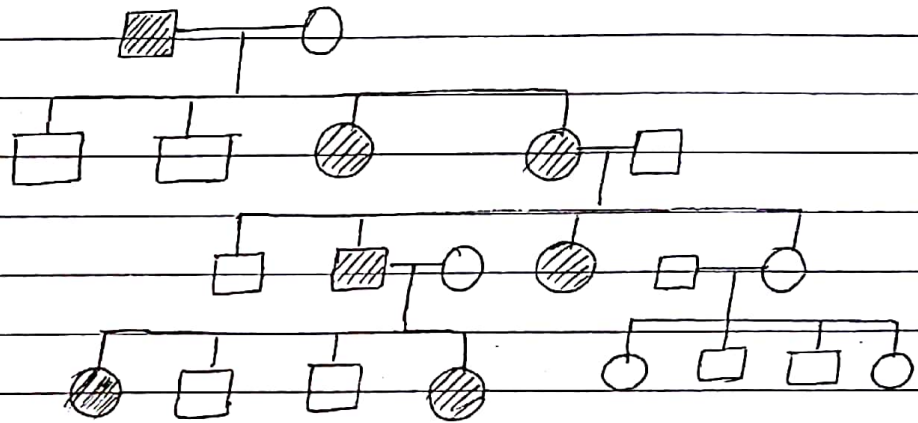
AR

↳ Cystinosis

- Metabolic - Galactosemia, HF-1

Tyrosinemia (R-Nitisinone).

- Acquired - expired tetracycline



X-linked dominant father to
all daughters - none sons

X-linked dominant: Males are more severely affected

- Familial hypophosphatemic rickets.
- Urea cycle defect due to OTC deficiency.
- Incontinentia pigmenti
- Rett's syndrome.

↓
bcoz boys die.

(lethal in male fetus).



Rett's Syndrome:

- Pervasive ~~person~~ developmental disorders.

↳ Autism (Common in boy) < 3yr

Asperger

Rett's

Q.

Not seen in autistic disorder -

A) Social avoidance

B) Visual impairment.

C) Interest in one self

D) Introvert person.

Asperger's Syndrome: Common in boys

Very good IQ.

Rett's Syndrome - Common in girls

X-linked dominant.

Normal till 6-18 months

Decrease in head growth.

Microcephaly; MR

Hand movements

Repetative behaviour

MECP2 gene mutation

↳ Macrocephaly not seen.

↳ Abnormal dendritic morphology in cortical pyramidal cells
(postmortem brain biopsy).

↳ Seizures.

Cause of death → Arrhythmias (Cardiac)



Sudden death.



Ornithine Transcarbamoylase Deficiency:

Orotic aciduria → URACIL

→ X-linked disorder.

- The mother of these child have also high Uracil level in urine.

Deletions:

Major deletion

→ Cri-du-chat

↳ Deletion of 5p chr.

- Cry like cat due to abn larynx.

Microdeletion

→ Williams (7q23-) Syndrome

→ Prader-willi Syndrome (15q11-13-)

→ Di-George's Syndrome (22q11-)

Δ → by FISH.

William's Syndrome:

Supravalvular aortic stenosis

↳ peripheral pulm. stenosis.

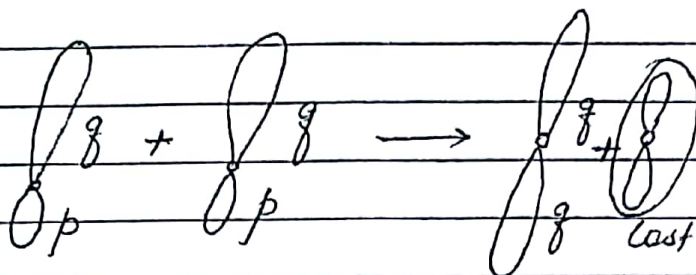
- Hypercalcemia
- Elf in facies.

Di-George's Syndrome:

- Hypoplasia of 3rd & 4th parapharyngeal pouch.
- Absent $\left\{ \begin{array}{l} \text{Thymus} \\ \text{Parathyroid.} \end{array} \right.$



TRANSLOCATION:



- Unbalanced

- Robertsonian Translocation:

Translocation b/w two Acrocentric Chromosome.

M/c/c (Genetic) of MR = Trisomy 21 (Down's Syndrome)

Extra chromosome is of females.

Trisomy 21 - In 95% cases: Maternal meiotic non-dysjunction.

- 3-4% - Robertsonian translocation

- 1-2% Mosaic 47/46

carrier of RT look normal but they can have abn children.

MOSAICISM:

A single zygote giving rise to different cells.

CHIMERA - different zygote giving rise to different cells
↳ Rare in humans.

- Mosaicism Seen in humans

- 1-2% are down's

Klinefelter Syndrome → 80% XX Y; XY/XXY;
XY/XXXXY.



Turner's Syndrome:

Cytogenetics

60% 45XO

15% Mosaic XX/XO

10% Isochromosome Xq or Xp. — Mentally Retarded.

10% 46X deletion.

5% Mosaic XO/XY → Risk of Gonadoblastoma.

Loss of one arm & duplication of other



Isochromosome.

Mosaicism { Somatic — Not transmitted

Germline — Transmitted

↳ Blood DNA is normal.

eg: ~~Germline~~ Osteogenic imperfecta.

Q.

Couple has two children c/ tuberous sclerosis. On detailed clinical & lab evaluation (including molecular studies) both parents are normal. ≤ one of the following explains the 2 affected children in this family —

A)

B)

C)

☒ D) Germline mosaicism.

#

Maternal inheritance → Mitochondrial.

Mitochondrial inheritance:

- MERRF (Myoclonic epilepsy & ragged fibres).
- Mitochondrial encephalopathy, stroke-like episodes, & lactic acidosis (MELAS)
- Leber hereditary optic neuropathy (LHON)



- Leigh disease
- Kearns - Sayre Syndrome (KSS) (ophthalmoplegia)
- NARP (Neuropathy, Ataxia, Retinitis pigmentosa)
- Chronic progressive ophthalmoplegia
- Pearson's Syndrome: Pancytopenia
+ Pancreatic insufficiency

Anticipation: Severity of genetic disorder \uparrow \bar{c} every successive generation.

More repeats — more problem.

eg: All Trinucleotide repeat disorders

- Fragile X — CGG repeats
- F. Ataxia — GAA "
- Myotonic dystrophy — CTG, CCTG
- Spinobulbar muscular atrophy — CAG
- Huntington's — CAG
- Spinocerebellar ataxia — CAG/CTG.

Fragile X: genetic

- 2nd m/c cause of MR in boys.
- X-linked
- Large face
- Large ears
- Prominent jaw
- Large testis.
- CGG repeats > 1500 repeats

Chr. 15q - 11-13 deletion

↳ Material "Angermann"

- Differential expression of genetic appearance depending upon parents.

Prader-willi Syndrome

- Severe neonatal hypotonia.
- Obesity
- ~~Small~~ Small hands & feet. (Rx - GH treatment)
- Unusual behaviour
- MR.
- High Ghrelin \rightarrow So obese. (Rx - Anti Ghrelin)

Angelman Syndrome:

- (N) at birth
- k/A happy puppets.
- Subsequently develop seizures
- MR
- Ataxia

↓
appetite hormone
for fundus.

Genomic imprinting → Prader Willi Syndrome (70%)

Almond shaped eyes.

- ② Angelman Syndrome (70%)
- ③ Neonatal DM
- ④ Beckwith Weidman
- ⑤ Seckle
- ⑥ Temple
- ⑦ Wang
- ⑧ Pseudohypoparathyroid Ib.



Q. Father carrier of cystic fibrosis - AA^c
Mother - $(N) \rightarrow AA$



Yes there is a chance of cystic fibrosis in child

AA^c



Uniparental disomy.

- In some cystic fibrosis
- In some sickle cells.

- 30% Prader Willi - Unimaterial disomy.
- 5% Angelmann - Uniparental disomy.

PRADER WILLI :

- 70% - Paternal inheritance deletion.
- 30% - Unimaterial disomy
- ↳ Maternal silencing gene.



Congenital Heart ds:

NADA'S CRITERIA:

1 Major & two minor criteria at least.

Major:

- ① Systolic murmur grade III or more ass. \bar{c} thrill.
- ② Diastolic murmur
- ③ Cyanosis (Central)
- ④ CHF

Minor:

- ① Systolic murmur < grade III
- ② Abn S_2
- ③ Abn ECG
- ④ Abn X-Ray
- ⑤ Abn BP

A_2 ----- P_2 Expire
 A_2 ----- P_2 Inspire

ASD - Wide & fixed S_2 .

A_2 ----- P_2 Expire
 A_2 ----- P_2 Inspire.

- Volume overload in right ventricle.

VSD - Wide & Variable S_2 .

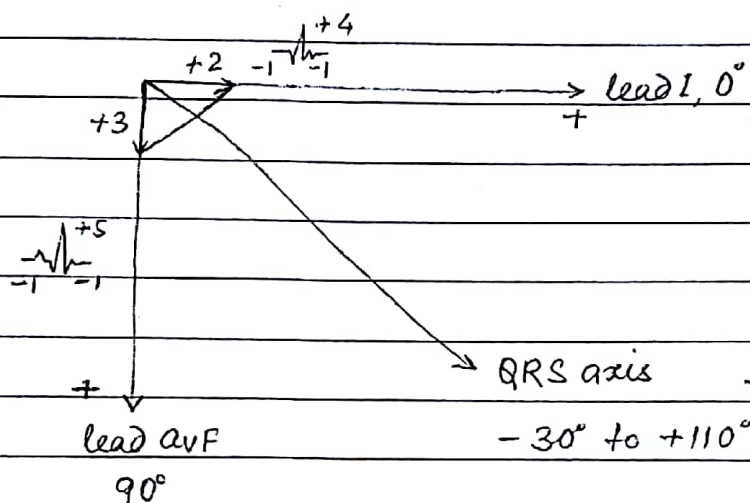
A_2 ----- P_2
 A_2 ----- P_2



TOF - Single S_2 (A_2).

All newborn has RVH & RAD.

- Axis like adult > 1st month life.
- T-wave V_1 ; V_3R ; V_4R
- 'up' first 48 hrs.



- Negative after 48 hrs
- Never be +ve < 6 yrs
- > 6 yrs → positive.

Prevalance of CHD:

- Prevalence = 0.8 - 1%

Recurrence = 2 - 6%

M/C CHD = VSD (30 - 35%).

2nd M/C CHD = ASD (Secundum) + 6 - 8%

> PDA (6 - 8%)

> Co-arcuation of Aorta (5 - 7%)

> TOF (5 - 7%)

> Pulm. Valve stenosis (5 - 7%)

> Aortic " " (4 - 7%)



M/C Syndrome in CHD = Down's Syndrome.

Down Syndrome:

M/C → Complete AV septal defect (CAVSD) /
AV canal defect / Endocardial cushion defect
Ostium primum ASD.

(37%) - M/C/c of death.

- VSD (31%)
- ASD (15%) → Secundum
- Partial AV septal defect (PAVSD) - 6%
- TOF → 5%
- PDA → 4%
- Miscellaneous → 2%

Turner's → half to One third bicuspid aortic valve;
20% coarctation.

Noonan's → Turner's phenotype.

AD; XX = XY

Valvular pulm. stenosis.

HOCM; ASD.

Rubella → PDA; Peripheral pulm. stenosis; VSD.

↓
Rubella

Alagille Syndrome

William's "

Maternal lithium → Ebstein's anomaly.

Maternal mumps → Endocardial fibroelastosis/
LV obstruction newborn.

Maternal penicillamine → Cutis Laxa.



Maternal SLE << Maternal Sjogren Syndrome

- New born complete heart block.

pacemaker insertion (anti ~~flow~~ Ro).

Maternal warfarin:

Chondrodysplasia punctata.

Maternal Thalidomide: Phocomelia.

Foetal circulation:

← 1 Umbilical Vein (left)
2 umbilical artery.

Ductus venosus →

PO_2 in umbilical vein = 30-35 mm of Hg

PO_2 of IVC in fetus = 28-30 mm of Hg.

As soon as child takes his 1st ~~breath~~ breath

- Umbilical artery constricts.



U. vein closes



Ductus venosus closes.

Ductus arteriosus:

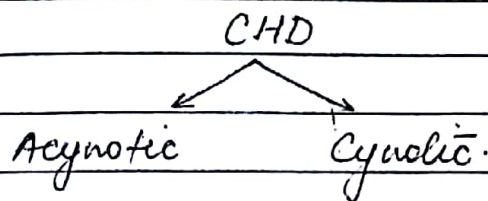
- Physiological \bar{c} in 10-15 hrs.

- Anatomical → 10-21 days.

Foramen ovale:

Functionally closes by 3 months.

Anatomically 10-15% open.

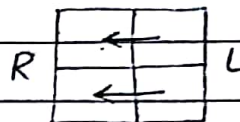


Acyanotic: L → R Shunt

ASD, VSD, PDA.

Pulm. blood flow ↑ (Plethora)

↳ lung vascularity is good.



CF: Failure to thrive

Recurrent pneumonia

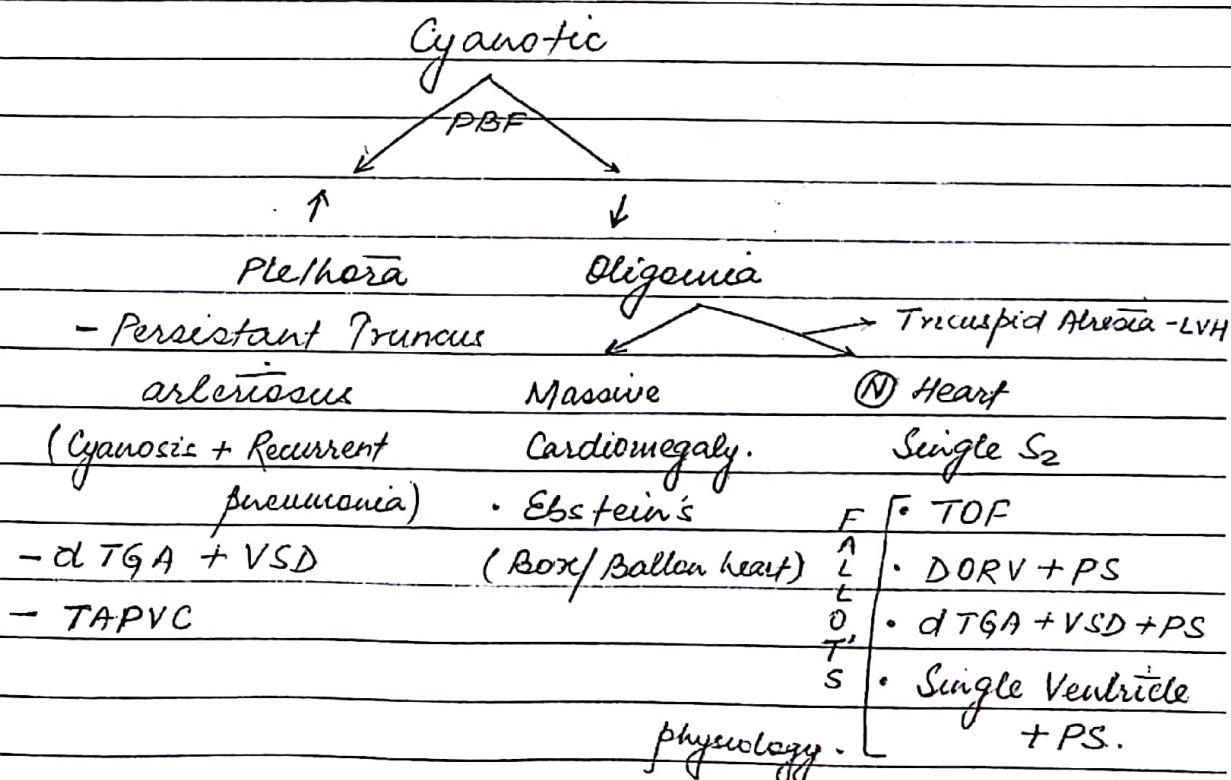
Feeding diaphoresis

CHF in 6-10 wks of life.

Suck-rest-suck cycle

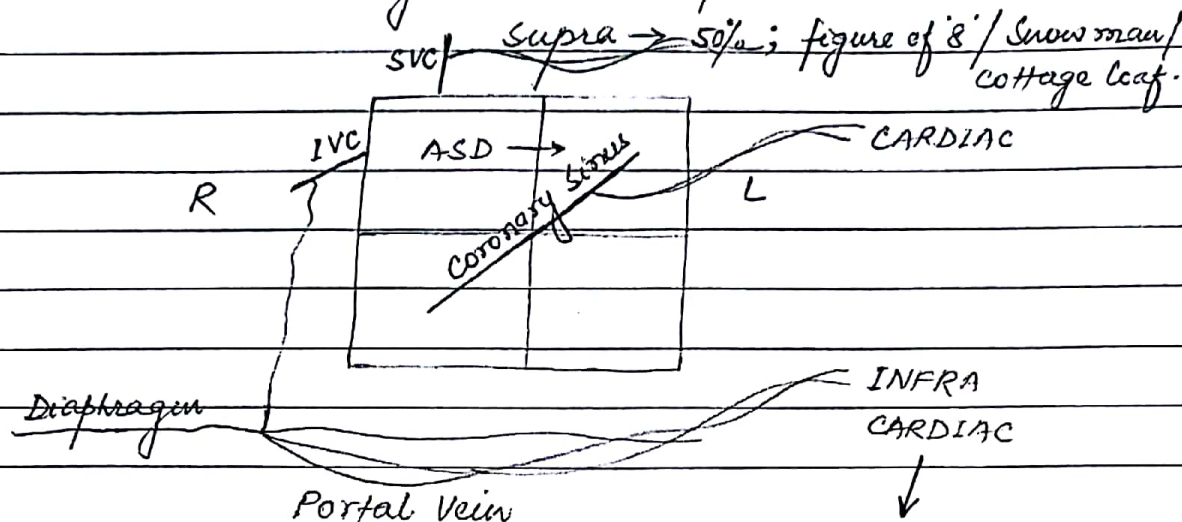
↑ sweating (d/t sympathetic stimulation).

Tachycardia
Cardiomegaly] in every CHD.





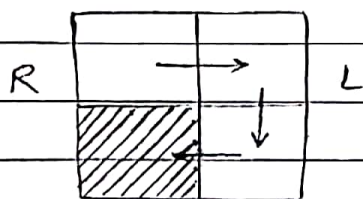
TAPVC (Totally anomalous pulm. venous connection):



More severe TAPVC.

- Cyanosis at birth.
- Obstructive pulm. venous hypertension.
- Ground glass: Kerley B lines.
- ⊖ - Worse by PGE1 infusion.
- Only pediatric cardiac Sx emergency.

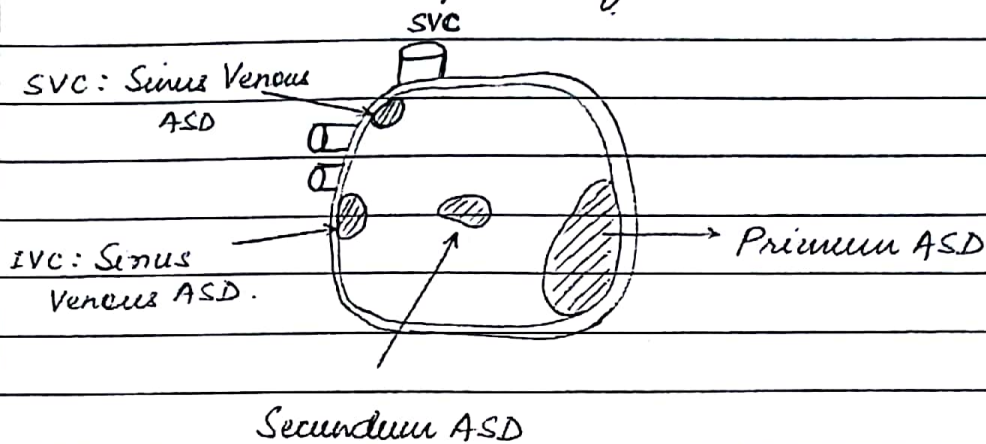
~~Tricuspid Atresia~~ Tricuspid Atresia:



- Cyanosis + ↓ PBF
- +
- LVH
- LAD

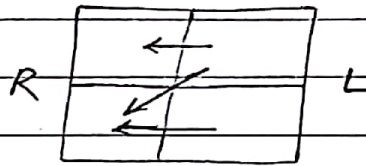


ASD (Atrial Septal defect):



M/C syndrome ass. = Down's Syndrome.

Endocardial cushion defect.



- Very large L-R shunt.
- PBF \uparrow
- Pulm. plethora

ASD Syndrome:

- HOLT ORAM : • Familial, AD, heart
- ASD Secundum; VSD; 1 degree block.
- AF + Bone defect (Absent Radius).
- Distally placed thumb / Rudimentary thumb / Triphalangeal thumb.
- A/K/A Hand-heart Syndrome.
- TBX5 mutation \rightarrow Pleiotropy.
 \hookrightarrow Common transcriptional factor for hand & heart.



Absent Radius associations:

- ECG (Holt-Oram)
- Platelet (Thrombocytopenia; AR)
- Bone marrow biopsy (Fanconi's anemia)



Congenital aplastic anemia

- Rarely Karyotyping (Edward Syndrome)

ASD Syndromes:

- Down's Syndrome
- Holt Oram
- Luteimbacher → ASD + Mitral Stenosis
- Ellis Van Creveld → ASD + polydactyly.

ASD Secundum:

- Child → asymptomatic, wide & fixed S₂.
- ECG → RAD (Right axis deviation).
- In adult life → Complications



- RV failure
- Arrhythmias; AF → CVA
- Reversal; R → L (Eisenmenger Syndrome)

Natural history:

- If < 3mm → close itself
- > 8mm → Unlikely to close; Require Sx.

Indication of Sx in ASD Secundum:

- All symptomatic.
- Q_p/Q_s > 2; even if no symptoms.
↳ Pulm blood flow / Systemic blood flow.

Q.

Least chances of Infective endocarditis is seen in ?

- A) Small VSD \rightarrow M/c/c of IE
- ☒ B) Small ASD secundum \rightarrow Rare
- C) Mild AS
- D) Mild AR.

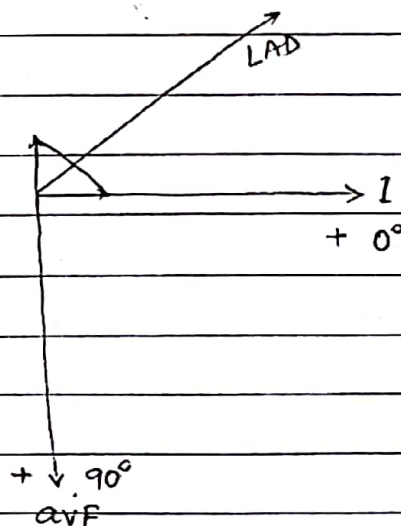
ASD Secundum doesn't require any prophylaxis t/t before going to Sx.

ASD primum + Mitral Regurgitation:

- Wide & fixed S_2 +
 $S_1 = = = S_2$ apex \rightarrow axilla & back.
- 6-10 wks presents \bar{c} CHF.
- Conduction defects; ECG \rightarrow LAD.
- Common in Down's Syndrome baby.

\downarrow
LAD d/t endocardial Cushion defect.

Down's
AVSD





VSD (Ventricular Septal defect):
70% → Perimembranous

Small VSD

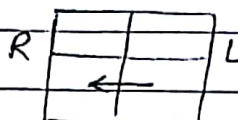
- Root of aorta $< \frac{1}{3}$
- $< 3 \text{ mm}$

- Called Maladie de Roger's defect.
- Loud murmur (S_1) (Pansystolic)

↓
Lower left Sternal border.

- Asymptomatic

Medium VSD



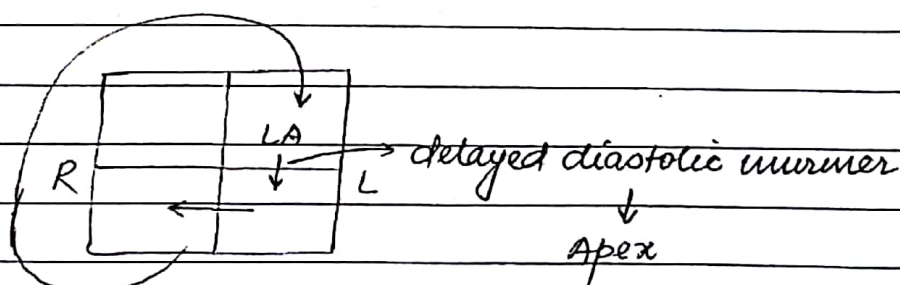
← CHF at 6-10 wks of life →

Large VSD

- $> \frac{1}{2}$

- No murmur/ Ejection systolic murmur.
- (b/w S_1 & S_2).

In medium to large VSD, Left Atrium enlarges first.
In Small to medium VSD, left Ventricular hypertrophy.
due to blood overload.



Natural course of VSD closure:

- 80% perimembranous
- 50% muscular VSD's close.
- By 4 yrs.



Indication of Sx in VSD:

- Failure of Medical therapy.
 - ↳ Digoxin
 - ↳ Diuretics
 - ↳ Dilators
 - ↳ ACE i (Remodelling of heart).
- $Q_p/Q_s > 2$; ^{even} if no symptoms.
- Swiss cheese VSD. (Multiple; Apex)
- Supracristal (outflow).

Large L → R shunt

↓
Pulm. blood flow ↑

↓
Irreversible changes pulm. microvascular.
[Pulm. HTN]

↓
RV pressure ↑; RVH

↓
Reversal; R → L (Eisenmenger Syndrome).

⇓
Sx is C/I!

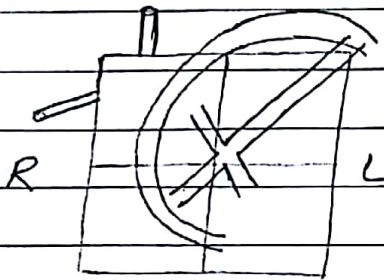
Differential clubbing: Toes > fingers

↓
PDA + Reversal.
(also differential Cyanosis).

Down's Syndrome baby c Endocardial cushion defect
undergoes Eisenmenger Syndrome.

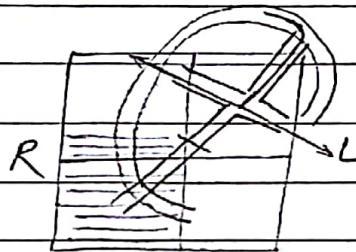


Ductal dependent lesion:



- HLHS \Rightarrow R - PGE₁ infusion. \nearrow S/E - Apnoea.
- Critical AS
- Preductal coarctation
- Interrupted aorta
 \hookrightarrow Shock.

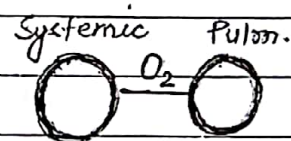
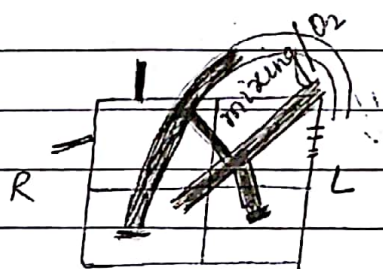
- I> Systemic blood flow dependence.
- II> Pulm. blood flow dependence.



- Tricuspid Atresia
- Pulm. Atresia
 \downarrow
Central cyanosis.

In emergency condⁿ \rightarrow PGE₁ infusion.

III> d-TGA



R - PGE₁ infusion

d-TGA \rightarrow dependent for mixing.

Truncus Arteriosus is Ductal independent.

Emergency Sx in TGA \rightarrow Rashkind's Balloon atrial septostomy.

Definitive Sx \rightarrow Jatene's Arterial Switch.
 \hookrightarrow Best time: \bar{c} in first 2 weeks of life.



M/c Cyanotic Heart ds:

Overall — TGA

In Infants — dTGA (<1yr)

> 1yr — TOF

Hyperoxia test in cyanotic newborns —

- 10L O₂ to cyanotic newborn
- pO₂ > 150 mm Hg → excludes heart ds.

① 50% d-TGA + VSD → Mild

② d-TGA + intact septa (complete d-TGA) — Birth.

③ d-TGA + patent foramen ovale → At birth.

① < ③ < ②

TOF (Tetralogy of Fallot):

M/c cyanotic heart ds beyond infancy.

- ① Narrowing of the pulm. valve (Infundibular).
- ② RV hypertrophy.
- ③ Overriding of aorta over VSD.
- ④ VSD — opening b/w left & right ventricles.
(perimembranous).

Pink child become a blue TOF.

- Cyanosis; Clubbing.
- Polycythemia → R/F of Renal vein thrombosis (RVT).
- Hematuria, ~~hematuria~~, Flank mass, Anemia.
- Infant — Cyanotic/tet/hyperpneic spells → older squats
- Complications:
 - ① < 2ys = Thrombosis



② > 2 yrs = Brain Abscess.

↓
in the territory of middle
meningeal artery
(Parieto-Temporal).

Infants \rightarrow Cyanotic/tet/hyperpneic spells — older squats
(R: knee chest position; — Systemic Vascular Resistance falls.

i.v. Ketamine; — R \rightarrow L Shunt \uparrow — Murmur disappears.

Phenylephrine) — PO_2 falls (R: O_2 ; i.v. $NaHCO_3$).

- Respiratory centre + + + (R: s/c Morphine)
- Dynamic Pulm. stenosis (R: i.v. Propranolol)
- Crying (Bcoz of \uparrow venous return).

In TOF — loud Holosystolic murmur at the left
2nd-3rd ICS due to pulm. stenosis.

Palliative Shunt

- Modified Blalock-Taussig \rightarrow Subclavian
to Pulm. artery
(opposite to Aortic Arch).
- Waterston
- Pott's (Descending Aorta to left pulm artery).



25% TOF \Rightarrow has Right aortic arch.

↓
M/c \bar{c} 50% of Truncus arteriosus

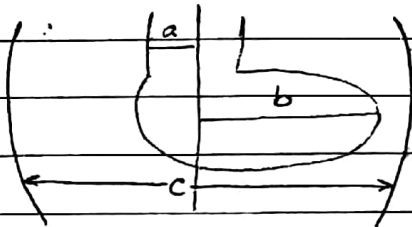


Pentalogy = + ASD

Trilogy = Overriding of aorta absent; VSD absent;
ASD present.

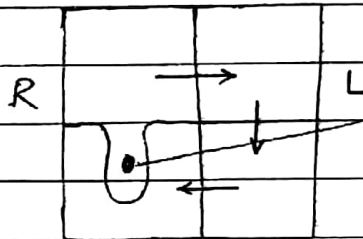
CHF never seen in TOF
Cardiomegaly never seen in TOF.

↓
Cardiothoracic ratio > 0.6 infant
 > 0.55 in older infant.



"Boot shaped heart".
"Coarcteur en Sabot"

Ebstein's anomaly:



- Cyanosis \downarrow PBF
- Maternal lithium

→ Atrialisation of RV.

Pressure = RA

Ventricular = ECG

disconcordance b/w pressure & ECG.

- Intracardial ECG helpful in Δ .
- Systolic; diastolic murmurs.
- Quadruple rhythm
- Box/ Ballon; Massive heart.
- SVT; WPW Syndrome.



Q A neonate present c recurrent abdominal pain; restlessness, irritability & diaphoresis on feeding. Cardiac auscultation reveals a non-specific murmur. He is believed to be at risk of MI. The most likely Δ is —

- A) VSD
- B) ASD
- C) TOF
- D) Anomalous origin of the left coronary artery = ALCA

- Q wave in Lead I; aVL
- LAD branch absent.
- Ischemic LV → Anterolateral MI.

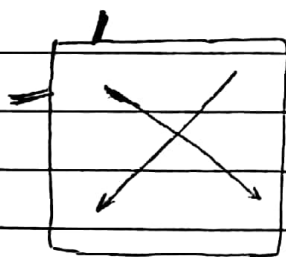
On Aortography → Rt coronary artery fills.
- Lateral MI in infant.

Rx: Bypass Sx.

L-TGA → Corrected TGA.

Normal Heart.

Atrio-Ventricular discordance.



[RA connected to LV
LA " " RV

No problem to baby.



KAWASAKI'S DS:

- M/c acquired heart ds in US, Japan, & Chandigarh.
- Fever > 5 days.
- Development of a limp.
- Erythematous macular exanthem over body.
- Ocular conjunctivitis.
- Dry & cracked lips.
- Red throat & cervical lymphadenopathy.
- Grade II/IV vibratory systolic ejection murmur at lower left sternal border.
- Predominant Neutrophils \bar{c} \uparrow platelet.
- M/c medium sized vasculitis.
- 20-25% causes develop aneurysm in future.

Q. Which of the following vasculitis not occur in adults?

- A) Kawasaki's (85% are < 5 yrs)
- B) Susac's Syndrome - Seen in adult females.
- C) Giant cell arteritis
- D) HSP

Peeling of palm & toes is classically seen in 2nd-3rd wk of life.

Rare ~~in~~ before 3 months of life.
DOC: IV Ig.

Δ : Fever > 5 days & any 4 of these 5

- ① Changes in extremities (eg: Erythema, edema, desquamation).
- ② B/L conjunctivitis (not ass. \bar{c} exudates)
- ③ Polymorphous rash (not vesicular)
- ④ Cervical lymphadenopathy.



- ⑤ Changes in lips & Oral cavity (eg: Pharyngeal erythema; dry/fissured or swollen lips, Strawberry tongue).

Non-classical feature of Kawasaki:

Arthralgia; Arthritis;

Thrombocytosis; Urethritis;

aseptic meningitis (Irritable)

M/c/c of death in Kawasaki —

Overall — Coronary artery aneurysm

Acute phase — Myocarditis.

Rx: Iv Ig 2g/kg in acute phase reduces risk 4-6%
Aspirin 100mg/kg/day X 2 weeks.

If Resistant to Iv Ig: 10-15% cases

↳ Add steroid (Methylprednisolone).

↳ Repeat Iv Ig

↳ TNF blockers → Infliximab; Etercept.

↳ IL-1 inhibitor → ANAKINRA

Recurrence Rate → 1-2% cases.

Mx of aneurysm:

Small → 50% resolve over 1-2 yrs.

Aspirin 3-5mg/kg/day.

Medium to large → Add Warfarin

Sx (may be bypass)



HSP (Henoch-Schönlein Purpura):

M/c vasculitis (small vessel) Overall.

M/c leucocytoclastic vasculitis.

- Palpable non-thrombocytopenic purpura.
- IgA, deposition of vessels in dermis.

Pathogenesis: Aberrant Galactosylation
Mesangio-proliferative disorder.

Small vessels → Skin

Arthritis

Arthralgia

GIT - mesenteric ischemia

Kidneys - HSP Nephritis (40-50%).



84% develop in 4 wks

91% " " 6 wks

97% " " 6 months.

- Microscopic hematuria

- Proteinuria

- 1-2% RPGN (over days to week).

↳ On Biopsy Crescent Seen

↳ ∴ Crescentic GN



proliferation of parietal
epithelial cells.

Rx: i.v. Methyl prednisolone.



Rheumatic Fever:

- M/c acquired heart ds in India / Developing country.
- Due to Group A β -hemolytic streptococci strains M-type 1, 3, 5, 6 & 18.
- Most frequent b/w 5-15 yrs.
- Latent period 3 weeks.
- Autoimmune: Molecular mimicry

Affects \leftarrow Myocardium
Muscle.

Modified Jones (2015):

- Era of Echocardiography \rightarrow Subclinical AR/MR.
- A/c to Risk area.

Low Risk — defined as having an ARF incidence < 2 per 100000 school-aged children (usually 5-14 yrs old) per year or an all age prevalence of RHD of ≤ 1 per 1000 population per year (Class IIa; level of evidence C).

Criteria = 2 major or 1 major + 2 minor
+ Essential criteria.

Recurrence = 2 major or 1 major + 2 minor
(OR)
3 minor



Major Criteria:

Low risk populations:

- Carditis (Clinical &/or Subclinical)
- Arthritis (Polyarthritis only).
- Chorea
- Erythema marginatum
- Subcutaneous nodules.

Moderate to high-risk populations:

- Carditis (Clinical &/or Subclinical)
- Arthritis
 - Monoarthritis or polyarthritis
 - Polyarthralgia (+)
- Chorea
- Erythema marginatum.
- Subcutaneous nodules.

Minor Criteria:

Low risk populations:

- Polyarthralgia
- Fever ($\geq 38.5^{\circ}\text{C}$)
- ESR ≥ 60 mm in the first hour &/or CRP ≥ 3.0 mg/dl.
- Prolonged PR interval, after accounting for age variability.

Moderate to High risk populations:

- Monoarthralgia
- Fever $\geq 38.5^{\circ}\text{C}$
- ESR ≥ 30 mm in first hour or CRP > 3 mg/dl.
- Prolonged PR interval - Also in MR & MS.



Rx:

- Aspirin 100mg/kg/day X 12 wks.
- Prednisolone X 12 wks.
 - 2mg/kg X 3wks → taper
 - Severe carditis or CHF.
- Crystalline Penicillin G X 10 days
- IE prophylaxis.

Mitral valve → M/c involved in RF

↳ Recurrence cause Mitral stenosis.



Primary prevention



In India 2^o prevention



Penicillin G

Benzathine (i.m.)

- 600,000 IU for children wt ≤ 60 lb

1.2 million IU for children wt > 60 lb

every 4 wks.

Or, Penicillin V (Oral)

↳ 250 mg twice a day

or, Sulfadiazine or Sulfisoxazole (oral)

↳ 0.5g once a day for pt. wt ≤ 60 lb

1.0g once a day for pt. wt > 60 lb.

For pt. allergic to penicillin & sulfa group.



Macrolides are given orally.



CATEGORY	DURATION
- Rheumatic Fever \bar{c} out Carditis	5yr or until 21 yrs of age whichever is longer.
- RF \bar{c} arditis but \bar{c} out residual heart ds (No valvular ds)	10yr or until 21 yr of age, whichever is longer.
- RF \bar{c} arditis & Residual heart ds (persistent Valvular ds)	10yrs or until 40yrs of age whichever is longer. Sometimes lifetime prophylaxis.

Q

- Blood pressure = 86/600 mm Hg
- 4 yr, unconsciousness.
- HR = 180/min
- CFT = 4sec

$\Delta = ? \rightarrow$ Compensated Shock.

Rx = 20ml/kg 0.9% NaCl.

SHOCK = BP < 10th centile for age & Sex.

Criteria for Shock:

Criteria for Hypotension by age.

Age	Systolic BP
Term neonates (0 to 28 days)	< 60 mm Hg
Infants (1-12 month)	< 70 mm Hg
Children (1-10 yrs)	< 70 + (age in yrs \times 2)
Children > 10yrs	< 90 mm Hg

Compensated Shock:

Rx - 20 ml/kg 0.9% NaCl

Repeat upto 60 ml/kg

\rightarrow CVP line (next step).

• low - fluid given.



- Normal → Cold epinephrin
- = Warm: Nor-epinephrin.

Hypertension BP > 95th centile

- Essential - 10% ; increasing
- Secondary HTN
 - ↳ Renal parenchymal - Reflux Nephropathy
VUR
 - ↳ Renovascular
 - Major → RAS ; RVT
 - Minor → HUS
 - ↳ Cardiac - post ductal coarctation (Turner)
 - ↳ Endocrinal -
 - Hyperthyroid
 - Cushing
 - Pheochromocytoma
 - CAH 11 beta / 17 alpha hydroxylase deficiency

End organ damage in HTN:

- Fundus
- Echocardiography concentric LVH.
- Any adolescent BP > 130/80
In children > 10 yrs → 120/80 → HTN
- Urine protein.

R_x: HTN

- Life style modification.
- Pharmacological therapy - end organ; Symptoms;
Severe > 99th centile.

↓
ACEi / ARBs.

ACEi C/I if GFR < 30 → causes hyperkalemia

↓
R_x → Amlodipine (CCBs)



Hypertensive Emergency:

- LVF \Rightarrow S3; Gallop; Basal crepts.
- Seizures.

Rx:

Best: i.v. Nicardipine infusion

Rx: Sodium Nitroprusside i.v. /

Esmolol /

Labetalol i.v.

Linezolid, Nitroprusside & Amphotericin B
should be covered.

Q.

A 12yr old boy c seizure.

BP = 200/140.

Femoral pulses not palpable.

$\Delta \approx ?$

A) Takayasu aortoarteritis (Rx - Prednisolone).

B) Grand Mal Seizures.

C) Fibromuscular dysplasia (FMD)

D) Renal parenchymal defect.



Tachyarrhythmias
- Based on QRS.

~~Wide~~

Wide > 0.09 sec (VT/VF)

- Pulseless

- Arrest.

Rx - Defibrillation 0.5-1 J/kg

- Stable

Rx - Lignocaine

Amiodarone.

Narrow

- Reentrant SVT

- HR > 220 infant; > 180 older

- P-wave absent; inverted

Vagal maneuvers

Stable

Rx - Adenosine (fastly
given c saline flush)



as close to the heart
as possible.

CHF; Shock

Rx - Synchronized Cardioversion

Respiratory

Cough & cold caused by Rhinovirus.

PMNCI G/L

Age = 2m - 12 months = RR \sim 50 or more.

Age 12m - 60m = RR \sim 40 or more.

• Chest is Indrawing = Pneumonia.
or fast breathing

Rx :- Give oral Amoxicillin for 5 day.

Any general danger sign \Rightarrow Severe Pneumonia.
or Stertor in calm child \Rightarrow or very severe dz

Rx

\rightarrow Give 1st dose of an appropriate A/b.
 \rightarrow Refer to hospital.

Q. 18 m old child weighing 11.5 kg comes to PHC.
with resp. difficulty. O/E lethargic RR = 46/min.
No chest retraction = Next Step.

Ans (B) Prescribe oral Antibiotic & Refer to higher centre.

\rightarrow Signs (< 2 months)

- Convulsions or
- Fast breathing (≥ 60)
- Severe Chest Indrawing.
- 10 or more skin pustules.

or a big boil or

If axillary temp 37.5°C

or above or less than 35.5°C

- or lethargic/unconscious
- less than 10 unit.

Next Step

Serious = Give i.v.

Bacterial \Rightarrow Gentamicin.
Sug[®]

\downarrow
Refer to
higher centre.

- Umbilicus red or draining → local
- Pus/Pus discharge from ear or <10 skin pustules → Back
- Give oral

- Causes of Pneumonia Age wise in India & world
- Neonates = Grp. B. streptococcus, S. coli.
<3 weeks.
- 3wk - 3m = Respiratory syncytial virus (RSV)
S. pneumoniae, H. influenza.
- 4m - 4y = RSV, S. pneumoniae, H. influenza.
- >5y = M. pneumoniae, S. pneumoniae.
- H. influenza → Vaccination → ↓ in incidence.

Viral pneumonia

- Prodrome.
- Diffuse, B/L
- Not lobar pneumonia
- Interstitial infiltrate

T/E: ① Doc for RSV = Ribavirin.

② Influenza = Amantadine, Rimantadine

Hins = Influenza A → Neuraminidase inhibitor.

↳ oral oseltamivir.

↳ Inhaled zanamivir.

Pandemic :-

Phase 1 - 3 Animals.

Phase 4 - May 27, 2009 (Mexico)

↳ Human-human transmission at community

Phase - 5 - 29/5/09 USA.

Human - Human transmission in one zone

Phase - 6 = June 11, 2009, US & India.

→ Human - human transmission in two zones

H₁N₁ Virus :-

- ss RNA.
- Belongs to Orthomyxoviridae.
- Size = 80-200 nm.

Types :- A, B, C.

Surface Ag = H (Hemagglutinin)
N (Neuraminidase)

As the reassortment occurs in swine flu.
= Swine flu.

∴ Antigenic shift & drift - No vaccines made.

Symptoms = Flu like.

(Complications of H₁N₁) → in High risk groups.

- ⇒ Pneumonia (viral)
- ⇒ Bacterial Superinfection.
- ⇒ ARDS like features.

High Risk groups :-

Comorbidity.

- They were associated with some ~~morbidity~~ comorbidity.
- Pregnancy
- Nephrotic syndrome, chronic illness, post transplant
- <1 yr., >65 yr.

Indications of oseltamivir

① Give it to all suspected cases of H₁N₁.

② Give it to all confirmed case.

Asis by throat swab & nasopharyngeal.

Swab & send for Real time PCR.

③ H₁N₁ influenza.

③ Give it to all household contacts, occupational
adults (post exposure in doctors)-

Dose = 75 mg Cap BD - 5d.

Prophylactic dose = 75 mg OD → 7-10 days -

Zanamivir

T/E dose = Two 5mg Inhalations (10mg total)
twice / day x 5 days.

Prophylaxis dose = two x 5mg Inhal[®] - OD.

Q: H₁N₁ 10 wks pregnant -

→ Give Osetamivir [Not teratogenic]
→ TE → only in late

Q: Pt on Rifampicin induces hepatic CYP450.

gets H₁N₁ Inf[®]

→ What happen to dose of Osetamivir

Ans = Remain same ~~the~~ Renal etc[®]

Q: Dose in haemodialysis

= Osetamivir dose is reduced.

Pneumonia

- St. pneumonia = Causes severe lobar pneumonia.

- Incub/ Id = 1-3 days

(Rx) - Ceftriaxone, DOC.

→ Resistant variety = DOC → Vanco + Ceftriaxone

(Vaccines) :- > 2 yrs. children = PPV - 23.

< 2 yrs = PCV - 7/11 = conjugated.

6, 10, 14 weeks, j/jb booster at 15-18 months.

[GAVI] :- Global Alliance for Vaccines & Immun[®]

- founded by Bill Gates & Melinda Gates
- Free vaccines in poor country

Staph. Aureus

- Max Mortality (10-30%)

Ch - Air filled cavity (Pneumatocele)
also seen in dk.

1) Klebsiella.

2) Kerosene oil poisoning

→ Pneumatocele can rupture & develop pneumothorax

• S. aureus = m/c of empyema in children.
(Plus in pleural cavity)

T/t = TCDT.

DOC for S. aureus = Cloracillin.

Vancomycin for MRSA.

• H. influenza = usually part of S. sepsis.
- Can have arthritis, meningitis.

Rx = ampicillin & chloramphenicol.
20-40% are resistant.

Rx: (Ceftriaxone) → DOC.

* Atypical pneumonia

→ Rare - <4ys - ~~2-5ys~~, >5ys.!

- Symptoms - dry cough.

- Interstitial pneumonia

- Organism = Mycoplasma Chlamydia.

Rx :- Macrolides.

15 month

Motor → Walks alone, crawls up stairs

Adaptive → Makes tower of 3 cubes, makes a line with crayon, inserts raisins in bottle

Language - Jargon; follows simple commands, may name a familiar object (eg. ball) responds to his/her name.

Social → Indicates some desires or needs by pointing, hugs parents.

18 month

Motor - Runs stiffly, sits on small chair, walks up stair with 1 hand held, explores drawers & waste baskets.

Adaptive → Makes tower of 4 cubes, imitates scribbling, imitates vertical strokes, dumps raisin from bottle

Language - 10 words (average), names pictures, identifies 1 or more parts of body

Social → Feeds self, seeks help when in trouble, may complain when wet or soiled, kisses parent with pucker

24 month

Motor → Runs well, walks up & down stairs, 1 step at a time, opens door, climbs on

furniture, jumps.

Adaptive → Makes tower of 7 cubes (6 at 2 months)
scribbles in circular pattern, imitates
H₂ stroke, folds paper once imitatively
Language → Puts 3 words together (subject, verb,
object)

Social → Handles spoon well, often tell about
immediate experiences, helps to undress,
listens to stories when shown pictures

30 months

Motor → Goes up stairs alternating feet

Adaptive → Makes tower of 9 cubes, makes V &
horizontal strokes, but generally will
not join them to make cross, imitates
circular stroke, forming closed figure

Language → Refers to self by pronoun I
Knows full name

Social → Helps put things away, pretends in
play

36 months

Motor → Rides tricycle, stands momentarily
on 1 foot

Adaptive → Makes tower of 10 cubes, imitates
construction of bridge of 3 cubes

copies circle, ~~imitates cross~~

Language - Knows age & sex, counts 3 objects correctly, repeats 3 numbers or a sentence of 6 syllables, most of speech intelligible to strangers.

Social → Plays simple games (in parallel with other children) helps in dressing (unbuttons clothing & puts on shoes) washes hands.

48 months

Motor → Hops on 1 foot, throws ball overhead, uses scissors to cut out pictures, climbs well

Adaptive → Copies bridges from parallel model, imitates construction of gate of 5 cubes, copies cross & square, draws man with 2-4 parts besides head, identifies longer of 2 lines.

Language - Counts 4 pennies accurately, tells story

Social → Play with several children, with beginning of social interaction & role-playing, goes to toilet alone

60 months.

Motor - Skips

Adaptive → Draws Δ from copy, names heavier of 2 weights

Language - Names 4 colours, repeats sentence of 10 syllables counts 10 pennies correctly

Social → Dresses & undresses, asks questions about meaning of words, engages in domestic role playing.



Fever & Stridor Inspiratory sound in upper airway.

Croup/LTB

- 75% parainfluenza
- Prodrome → Stridor

Barking Cough.

Subglottic narrowing

k/A = Stippel sign

On X-ray.

↳ Doc = formild = dexam = 0.6mg/kg.
wood & Suresh

↳ Nebulisation 2 Ralomic epinephrine.

↳ Westley Croup Score Criteria to dx it.

BRONCHIOLITIS:

- Inflammatory obstruction of smaller airway.

M/C organism - RSV (50%)

↳ Respiratory Syncytial Virus.

R/F:

- Males, Top fed.
- ① [- Preterm; Chronic lung disease.
- ② [- L → R shunts.
- Smoking mothers.

Airway resistance, $R = \frac{1}{r^4} \rightarrow$ air trapping

Prodrome → Wheeze / Ronchi.

X-Ray chest shows Hyperinflation.

Rx: Antibiotics has no role.

Humidified O₂.

For 2 High Risk group - Nebulised Ribavirin
Palivizumab



Long term - Persistent wheeze infancy

↓
Reactive airway disease.

ASTHMA

Classification a/c to severity.

	Step	Symptoms	Night symptoms	Peak expiratory flow rate.
I.	Intermittent.	< 1 time a week; asymptomatic & (N) PEFr b/w attacks	≤ 2 times a month	≥ 80% predicted; Variability < 20%.
II.	Mild persistent.	> 1 time a week but < 1 time a day	> 2 times a month	≥ 80% predicted; Variability 20-30%.
III.	Moderate persistent.	Daily use β -agonists; daily attacks affect activity.	> 1 times a month	> 60% & < 80% predicted; Variability > 30%.
IV	Severe persistent.	Continuous limited frequent activity.	Frequent.	≤ 60% predicted; Variability > 80%.

For intermittent → SOS β_2 -agonist

Mild persistent → +++ inhaled beclomethasone,
fluticasone, Budesonide.

Moderate persistent → +++ Salmeterol / Sustained release
theophylline.

Severe persistent → ++ Oral low dose, long term, alternate
day prednisolone.



2 yrs \rightarrow Acute severe asthma



Oxygen; Nebulise Salbutamol



Nebulised ipratropium bromide



i.v. hydrocortisone



s/c Terbutaline



Terbutaline infusion



50% MgSO₄ \rightarrow aminophylline.

Foreign body:

CxR \rightarrow Persistent inflation

Ball valve inflation.

Rx \rightarrow Bronchoscopy & remove foreign body.

Recurrent pneumonia:

2 episodes of radiographic pneumonia in 1 year

(OR)

3 episodes in any time frame.

Persistent pneumonia:

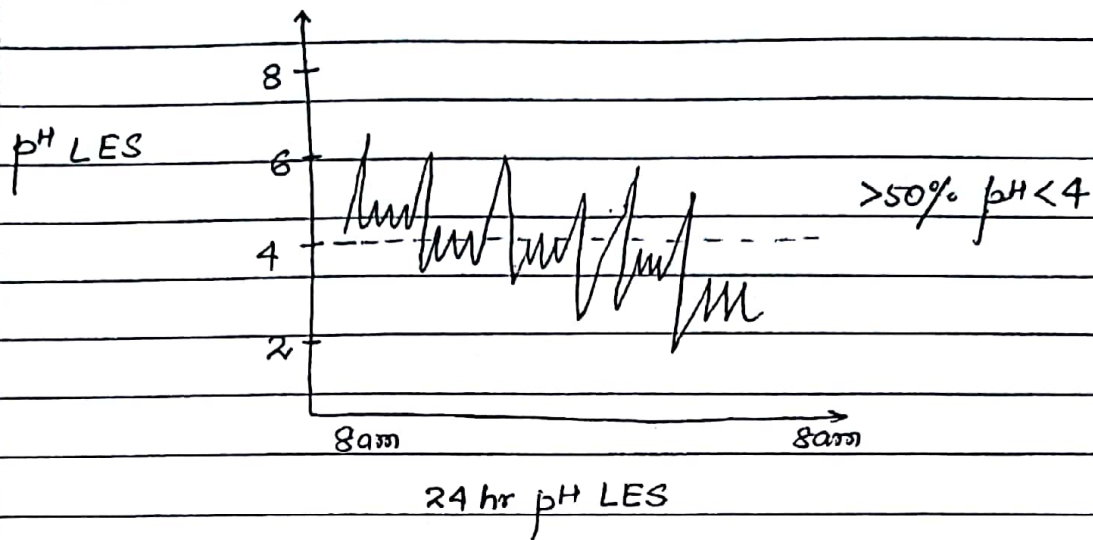
Persistence of symptoms & radiographic abnormalities for more than 1 month.

Recurrent pneumonia:

Cause \rightarrow L \rightarrow R shunt

GERD d/t aspiration.

Immunodeficiency.



M/c/c of Recurrent pneumonia in U.S. = Cystic Fibrosis.

Cystic Fibrosis


- Incidence: 1 in 2500 in UK

Gene → CFTR gene (7q 31.2 locus)

↓
cAMP regulated chloride channel.

- In CF more Sodium goes to lumen to mucosa than chloride.
- Autosomal Recessive disorder.
- Respiratory: Pneumonia (>5yrs of life).

M/c organism asso. c cystic fibrosis - *Pseudomonas aeruginosa*.
(Mucoid > Non Mucoid)

Rx:  Inhaled antibiotics

- Tobramycin
- Aztreonam
- Colistin
- Ciproflox



- Amikacin
- Levofloxacin

Respiratory: Pneumonia (>5yrs)

- <5yrs - S. Aureus; H. influenzae.
- 5-18yrs - S. Aureus; Pseudomonas
- >18yrs - Pseudomonas; S. aureus.

Acromonas; Achromobacter;
Burkholderia cepacia

- ↳ Specific; Fatal
- ↳ Rx Mild - Coltrimoxazole
- ↳ Severe - Meropenem +
Coltrimoxazole/Doxycycline.

- Exocrine Pancreas - Steatorrhea (<5yrs)
foul - bulky stools.



Rx - Steatorrhea in CF
Lipase 1000 IU/kg supplement.

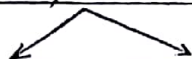
- Endocrine → 25% DM after 35yrs.

- GIT → Meconium ileum.

Adolescents - Distal intestinal obstruction Syndrome.

48hrs old baby has not passed meconium.

Ix → Lower GI contrast study



Diagnostic Therapeutic in meconium ileum.

- GIT: Diffuse pain abdomen
Colonic mucosal thickening.
Intussusceptions.
- Nasal polyps; Azoospermia



- Absent Vas : 1% infertile men ; common in CF males.

Q. Which glands are not obstructed in CF ?

- A) Cervix → Infertile
- B) Pancreas → Insufficiency.
- ✓ C) Sweat glands

M/c mutation in CF = $\Delta F508$ mutation (Class II mutation)
(Seen in 70% caucasians)
↓ (25-30% Indian)
M/c lethal genetic disorder in caucasians.

CFTR gene has 1800 mutations.

How many Nucleotide deleted in CF ?

3 → TTT = Phenylalanine.

Rx : For Trafficking Defect

↓
DOC { LUMACAFTOR (Trafficking corrector drug)
+
IVACAFTOR (Potentiator drug).

↓
opens CFTR 'Cl' channel.

Lumacaftor - induce hepatic Cyp450

↓
∴ New Corrector → TEZACAFTOR

↓
doesn't induce Cyp450.



Q Which of the following is a calcineurin inhibitor?

- ☒ A) Tacrolimus
- B) Sirolimus } mTOR inhibitor
- C) Everolimus }
- ☒ D) Cyclosporine

Diagnostic Criteria of CF:

- ① Sweat Chloride $> 60 \text{ meq/L}$ on 2 occasions
[② $< 40 \text{ meq/L}$]

OR, ② Two known CFTR mutations.

OR, Best - ③ Diagnostic nasal electrode potential difference.

Newborn screening for CF

IRT test \rightarrow Immuno reactive Trypsinogen
Assay test.
(Sensitive test)

Q Male, 10 month boy, Down Syndrome & Recurrent pneumonia

Filling defect \rightarrow In Ba-swallow

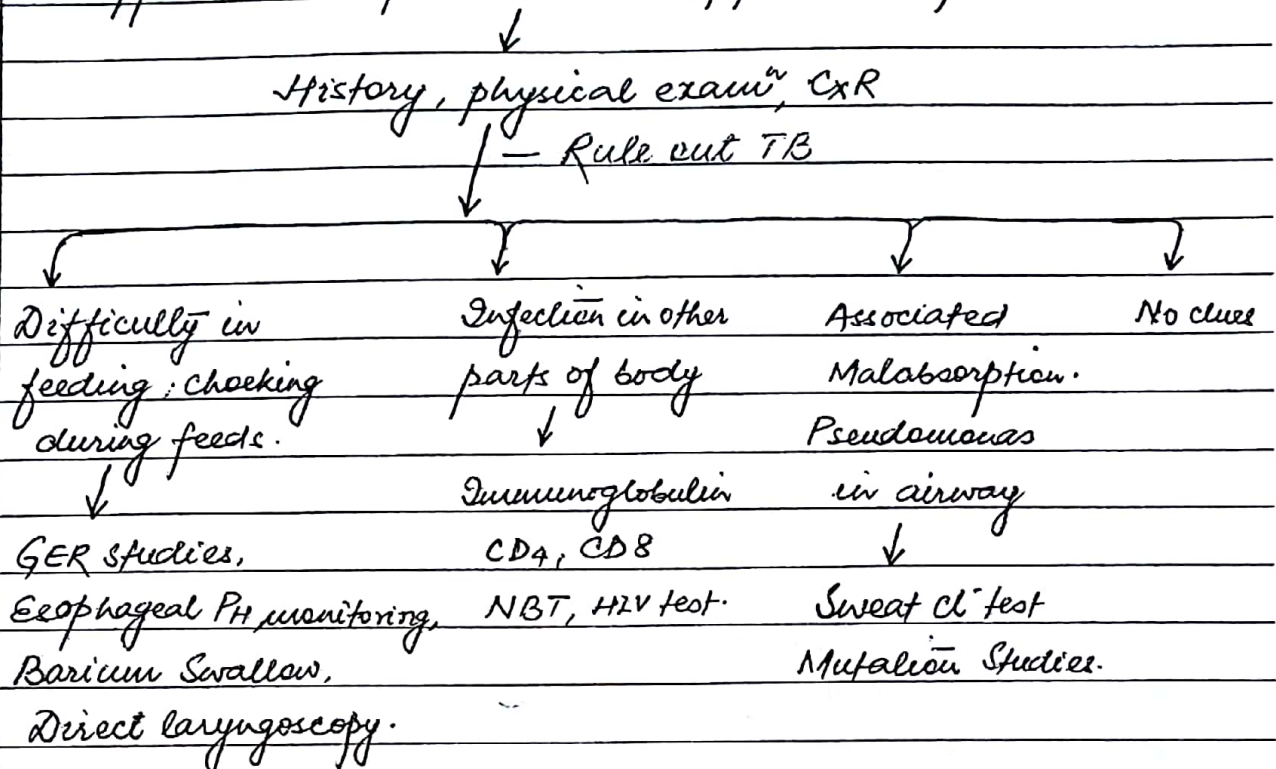
\hookrightarrow in middle of esophagus.

\hookrightarrow Abbarrent right subclavian artery
vascular ring

\downarrow
Dysphagia lusoria.



Approach to a pt. c recurrent / persistent pneumonia:



Isotonic fluids:

- 0.9% NaCl - 154 meq/L Na & Cl.
- Ringer lactate = Plasma.
 - 130 meq/L Na
 - 109 meq/L Cl
 - 28 meq/L lactate⁻
 - 4 meq/L K⁺
 - 3 meq/L Ca

Maintenance fluid:

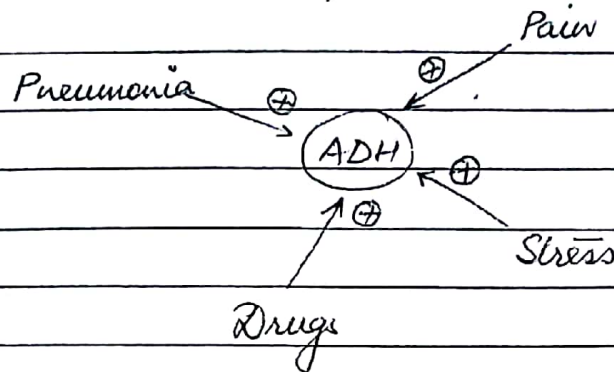
Need ?

- Insensible water loss.
- Energy
- To prevent catabolism.



Type ?

- Adult: 5% DNS
- Children 5% Dextrose + N/2 or NS.



Amount ?

A/c to Holiday & Segar

- < 10 kg - 100 ml/kg/day.
- 10 kg - 1000 ml/day. (40 ml/hr).
- 11-20 kg → 1000 ml (for 10 kg) + 50 ml/kg additional kg above 10 kg.
- > 20 kg → 1000 ml + 500 ml + 20 ml/kg additional above 20 kg.

Patau's Syndrome (Trisomy 13):

- Cleft lip & palate.
- Polydactyly
- Hypotelorism (Eye separated wide).
- Abortion looks like Cyclops
 - ↳ Holoprosencephaly.
 - (Single eye)
 - ↳ Fused frontal lobes + lateral ventricles.
 - ↳ Aplasia cutis (problem of cleavage of skin).
- ~~Rooster beak~~



Edward Syndrome (Trisomy 18)

- Rocker bottom feet
- Overlapping of fingers.
- 2nd M/c trisomy.
- Maternal age
- GIT anomaly are common
 - ↳ Atresia gut
 - ↳ Exomphalos
 - ↳ Malabsorption.

TTNB (Transient Tachypnea of New Born): Wet lungs.

R/F:

- Term ; Cesarean Section
- Macrosomia
- Precipitous labour.
- Maternal Sedation

CxR - Prominent horizontal fissure.

- It is a benign, self limiting condⁿ & resolves in 48-72 hrs.

FiO₂ requirement < 0.4

Never require mechanical ventilation.

MSL (Meconium Stained liquor):

- Marker of perinatal hypoxia (Common in Post term)

Glottis open

Parasympathetic ++

meconium passed.

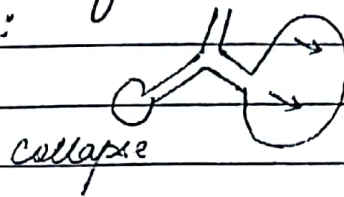
Peristalsis ++

Anal sphincter relax



Complication of Meconium:

Physical:



Ball valve.

air leaks 20-30%

Chemical: Irritant.

Biological \leftarrow impairs surfactant function (Surfactant given)
Good culture media (Given Antibiotic)

Rx:

Meconium stained liquor



Born

Vigorous

Zone.

HR > 100/min

Resp^r effort.

NO

YES

limp or,
HR < 100 or,
Apnea.

PPV X 100% O₂.

Transfer to mother



GROWTH

1-4 months : Weight gain 30gm/day

5-8 months : wt. gain @ 20gm/day

9-12 months : wt. gain @ 10gm/day.

Weight multiples:

Wt. $\times 2$ = 5 months

$\times 3$ = 1 yr

$\times 4$ = 2 yrs

$\times 5$ = 3 yrs

$\times 6$ = 5 yrs

$\times 7$ = 7 yrs

$\times 10$ = 10 yrs.

Length:

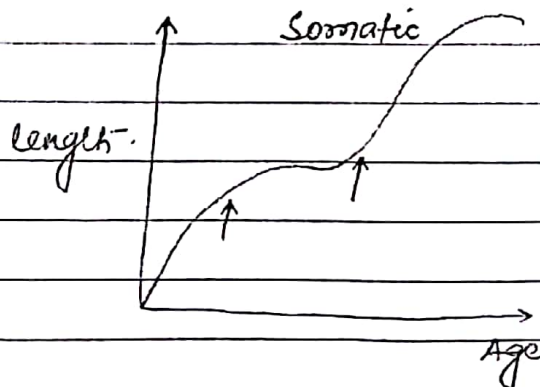
At birth - 50cm } Growth Velocity $\rightarrow +25$ (first year)

1 yr - 75cm

2 yr - 90cm $\rightarrow +15 \rightarrow$ second year.

4 $\frac{1}{2}$ yr - 100cm

Add 6cm / year till puberty.

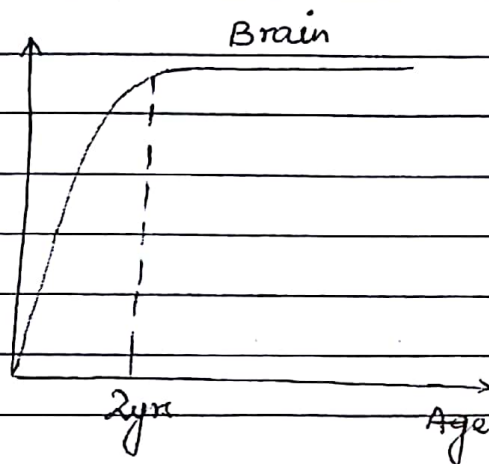


Q. In school going children; the avg height velocity is $\rightarrow 5-8$ cm

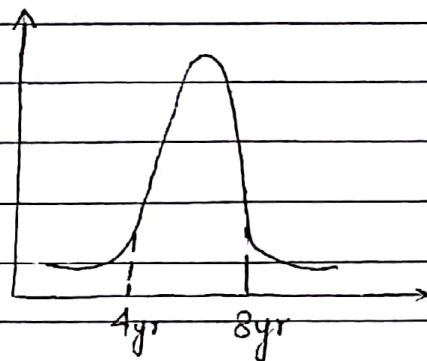


Head circumference:

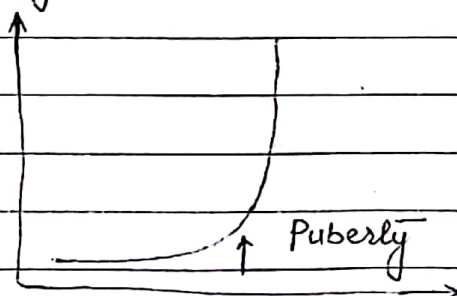
- 35cm at birth
- 3 months \rightarrow 40cm
- 12 months \rightarrow 45cm
- 2 yrs \rightarrow 48cm (90% of brain grows)
- 12 yrs \rightarrow 52cm.



Lymphoid growth:



Gonad growth:





Adolescent — 10 to 19 yrs.

Tanner's Sexual Maturity Rating :

SMR Stage I - V

Stage I — No character

Stage V — Completely developed.

Menarche → SMR-IV

Sequence of puberty:

Girls:

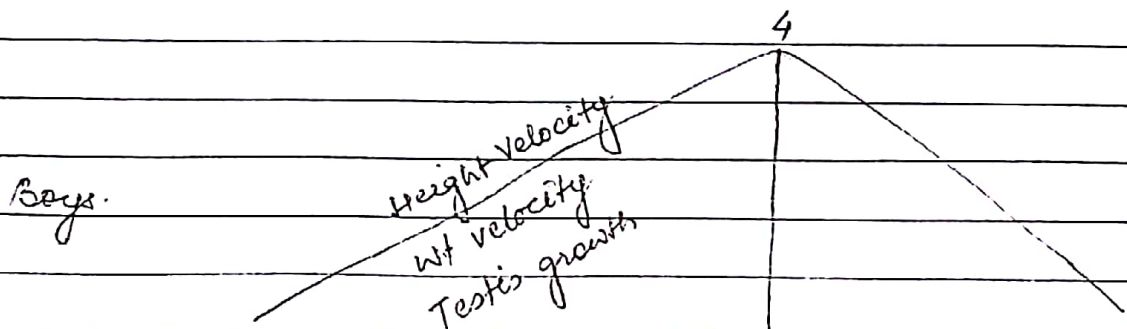
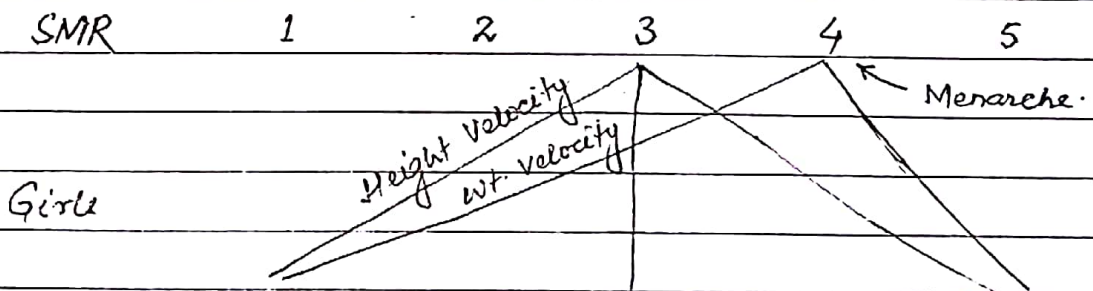
Growth Spurt → Thelarche, Pubarche, Menarche.

Boys:

→ Testis — Penis — Pubic hair, Axillary hair.

Girls : BP (Breast — Pubic hair)

Boys : GP (Genitalia — Pubic hair).



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Most rapid \uparrow in height \rightarrow Phase 3 in girls & Phase 4 in boys.

Puberty - comes by pulsatile release of GnRH.

Precocious Puberty:

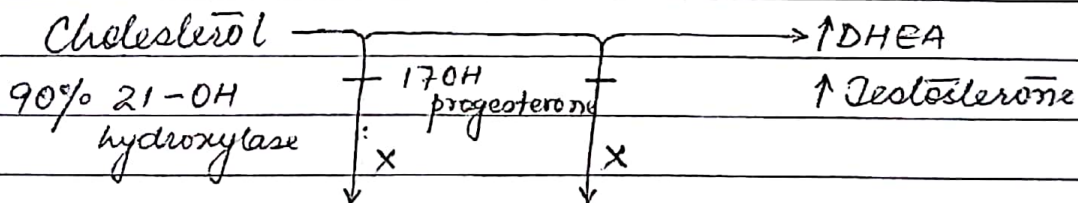
- Gonadotropin dependent / Central
- Females - Idiopathic.
- Males - Organic
(Hypothalamic Hamartoma,
Craniopharyngiomas,
Hydrocephalus, TBM).

CAH (Congenital Adrenal hyperplasia):

m/c/c of female hermaphrodite.

Karyotype - XX

But looks like Male.



Cortisol
glucose ↓

Aldos Cerone

$$Na^+ \downarrow$$

Salt-Wasting Crisis (70%)

 $\uparrow K^+$

Shock

- Cause of precocious puberty in boys.

Genitalia → Ambiguous

Labia is pigmented d/t ACTH.

- Virilization
- Hypertrophy of clitoris



- Penoscrotal hypospadias
+ Empty Scrotum.

Due to deficiency of 21-OH hydroxylase lead to accumulation of 17-OH progesterone.

Q Level of 17-OH progesterone in CAH

A) < 150

B) 150 - 300

C) 300 - 500

D) > 600 (> 3500)

CAH associated with Premature epiphyseal closure.



Short Stature.

Rx: CAH

- Supplement of Hydrocortisone & Fludrocortisone



Glucocorticoids

15-20 mg/m²/day

Thrice daily.



Mineralocorticoids.

0.15 mg/day.

- Girls require Sx

- Clitoroplasty

- Reconstruction Sx.

- Never get married.

Cholesterol

11 β -OH
deficiency

11-deoxycortisol

Cortisol

DOC: potent

Aldosterone

DHBA

Testosterone.

mineralocorticoid

HTN

Hypokalemia metabolic alkalosis.



Q. 5 yrs old boy has precocious puberty. BP = 130/80
Estimation of \leq help diagnosis?

- A) 17-OH progesterone
- ☒ B) 11-deoxy cortisol
- C) Aldosterone
- D) DOCA

Deficiency of 17 α -Hydroxylase \rightarrow \uparrow Aldosterone



HTN, Hypokalemia,
metabolic alkalosis.

\rightarrow X Cortisol

\downarrow glucose.

\rightarrow X Testosterone



Male look like ♀ .

(Mineralocorticoid) M

T (Testosterone).

α

\uparrow

\uparrow

\uparrow

\uparrow

\uparrow

3 β Hydroxysteroid dehydrogenase deficiency (3 β HSD def.)



Causes ambiguous genitalia in both sexes.

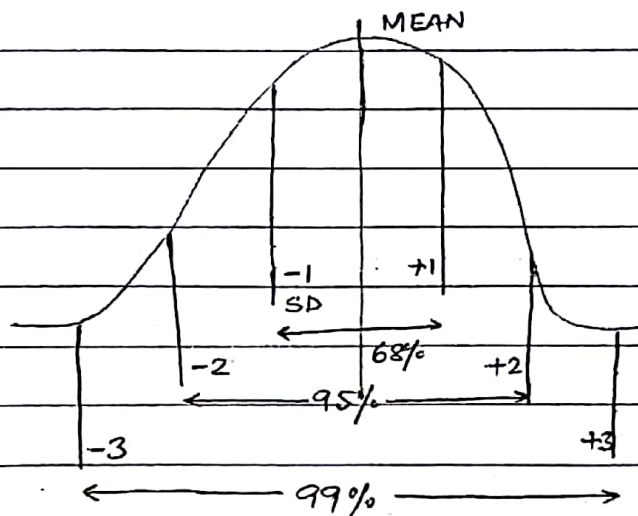


Q. Mother; previous child CAH

Next pregnancy - To prevent female virilization of foetus

Dexamethasone
(20mcg/kg pre-pregnancy wt.)

Inhibits ACTH.

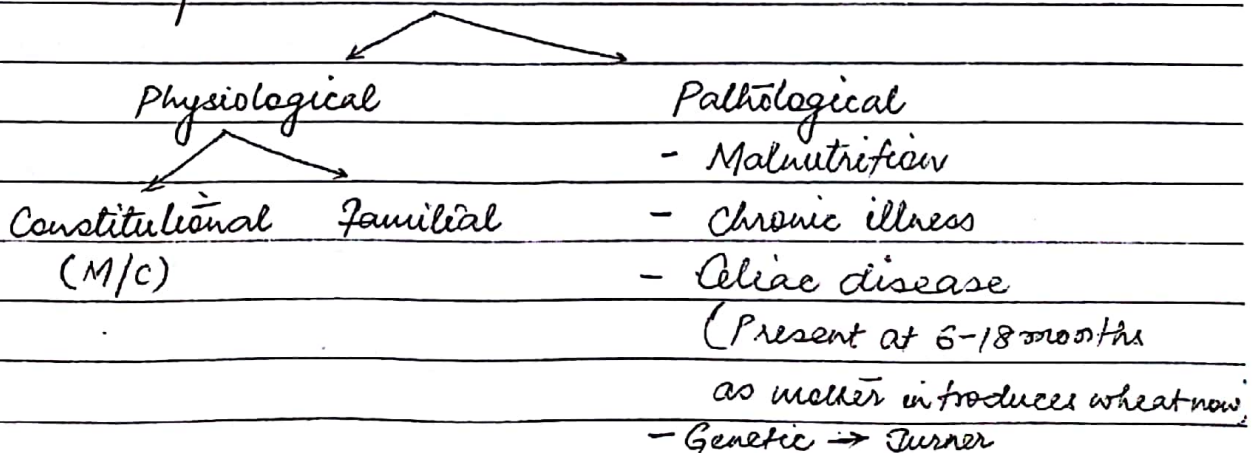


Short height > -2 SD below mean or $< 3rd$ percentile

Mid parental height = Adult predicted height

Boys = average parents height + 6.5cms
Girls = " " - 6.5cms.

Causes of Short stature:





Rx: Celiac ds

- Restrict Wheat, Rye & Barley for lifetime.
- Gluten-free diet.
- Also avoid oats.

↓

- Endocrinal → Hypothyroid
Cushing
CAH
GH deficiency.

GH deficiency:

- Birth weight & length normal.
 - Lag at 1-2 yrs of life.
 - Bone age delayed.
 - Doll face
 - Micropenis
 - Short stature
- } Panhypopituitarism

Micropenis (↓ LH, ↓ FSH)

↓ ACTH → Hypoglycemia

- Hoarseness of voice.

Inv: GH level ⇒ IGF, BP3

GH stimulation test - Best

↳ Basal level GH

↳ Stimulated level - 2 stimuli

(Clonidine; Insulin; L-arginine).

Rx - Recombinant GH (US, FDA)

Indication: rGH

- GH deficiency
- Turner Syndrome
- Prader villi
- Chronic kidney ds.
- SGA height - 2.25 SD below mean

When to stop GH?

- Height reaches 50th centile
- Epiphysis fuses.
- Pseudotumor cerebri
- Slipped capital femoral epiphysis.

Disproportionate short stature:

US/LS \uparrow \rightarrow Achondroplasia, Rickets;

Hypothyroidism:

US/LS ↓ → TB spine, Mucopolysaccharoidosis IV
(Morquio's ds)

Physiological Short stature:

	CONSTITUTIONAL (M/c)	FAMILIAL
Birth length	· (N)	IUGR
Lag	6-12 months	
Growth velocity	(N)	Less
Final Height	(N) / Sub (N)	Less
Puberty	Delayed	(N) [♀]
Bone age	Delayed.	(N) [♀]



Developmental milestones:

Neck holding → @ 3 month.



Gross motor milestones:

3 months - Head holding; Neck holding.

5 months - Sitting \bar{c} support.

4-6 months - Prone to supine, supine to prone
(Roll in bed).

8 months - Sitting \bar{c} out support.

9 months - Crawling

10 " - Creeping; stand \bar{c} support

12 " - Standing \bar{c} out support; walking \bar{c} out support.

2 yr - Walk up stairs \bar{c} two feet at each step.

3 yr - Upstairs \bar{c} one foot at each step, rides tricycle.

4 yr - Hops on one foot

5 yr - Skips on two foot

Fine motor:

12 wks \rightarrow Moro's reflex disappears

Grasp reflex disappears

4 months - Goes for objects

5 months - Bilateral grasp.

6-7 " - Transfer object, palmar grasp.

9 " - Pincer grasp; mature neat.

13 months - Casting.

15 " - Self feed \bar{c} a spoon

18 " - Self feed \bar{c} a cup.

24 " - turns pages of a book one at time.



Social Milestone:

- 2 months — Social smile
- 3 " — Recognizes mother.
- 6-7 " — Smiles at mirror image.
- 9 months — Waves bye-bye.
- 6 months — Stranger anxiety.
- 2yrs — Dry by day
- 3yrs — Dry by night
- Dress / undress himself (Supervision)
- ↓
- Out supervision — 5yrs.

18 month — Separation anxiety / Clinginess / Reproachment.

Language Milestones:

- 1 month → Head turns to sound
- 3 months → Cooing
- 6 months → Babbling; Monosyllables (ma, ba)
- 9 months → Bisyllables (mama; ba-ba)
- 1 yr → 2 words w meaning
- 18 months → Vocabulary of ten words.
- 2yrs → Simple sentences w 2 words; Phrase.
- 3yrs → tells age & sex; uses pronouns, handedness; identify colours.
- 4yrs → tells story
- 5yrs → Knows colour
- 15 months : 4-5 words
- 2yrs → 50-100 words



When can a child understand death =

< 3 yrs - No idea

3-9 yrs - Idea

> 9 yrs - Entity; Irreversible; himself

Object permanence/Constancy - 9 months.

Cross a busy road - 10-12 yrs.

Tie shoe lace - 5 yrs.

Bladder control - 85% by 5 yrs.

Nocturnal enuresis - > 5 yrs.

↳ more than 2/week x 3 months.

Q. 14 yrs old child c Nocturnal enuresis

Rx - • Behavioural - +ve reinforcement.*

• Alarm therapy (Best)

↳ 85% relapse free rates.

• Drugs → Desmopressin

Encopresis : > 4 yrs

Cause → Chronic constipation.



CNS

MICROCEPHALY:

→ HC > -3SD below mean

Causes: GENETIC

- Trisomy 13, 18, 21
- Cri-du-chat Syndrome

ACQUIRED

- Baby: HIE; Hypoglycemia, PKU, Meningitis, encephalitis.
- Mother → TORCH, ^{phenylalanine} Hyperphenylalaninemia > 6mg/dl, Alcohol, DM, Radiation.

MACROCEPHALY

HC > 2SD above mean.

Q Neurodegenerative disorders:

GREY MATTER:

- Normal at birth.
- Regression milestones.
↳ Disappear as they come orderly.
- Deaf, blind, seizures.
- Anemia, Hepatosplenomegaly.
- Cherry red spot macula.

Microcephaly

- GM1 gangliosidosis.

- Gaucher's ds

(β -gluco

- Niemann Pick's ds

(

Macrocephaly

- GM2 gangliosidosis

(Tay Sachs' ds)

- Sandhoff

Huge
spleen

VLCFA = Very long chain fatty acid.



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Jay Sachs ds -

- AR
- D/t deficiency of β -hexose aminidase A.
- 1/25 askenazi jew are carrier.
- 6 months exaggerated startle reflex.
- Cherry red spot macule
- Organomegaly not seen.

Organomegaly + Jay Sachs = Sandhoff



Def. of β -hexose aminidase A & B.

WHITE MATTER DISORDERS:

- All the tracts are white matter.
- Frequent fall, incoordination
- Upper motor neuron sign.

Microcephaly / (N)

- Krabbe's
- XLR adrenoleukodystrophy.



degeneration starts from
parieto-occipital area.

$\Delta \rightarrow \uparrow$ VLCFA levels

Rx \rightarrow Early bone marrow Tx.

lorenzo oil

- Metachromatic

leukodystrophy

Macrocephaly

- Canavan ds.
- Alexander ds.
- MRI \rightarrow Diffuse white matter thickening.
- MRI \rightarrow Degeneration starts from frontal periventricular area.



Hydrocephalus:

- Enlarged ventricles \bar{c} or \bar{c} out \uparrow in ICT.

CSF production:

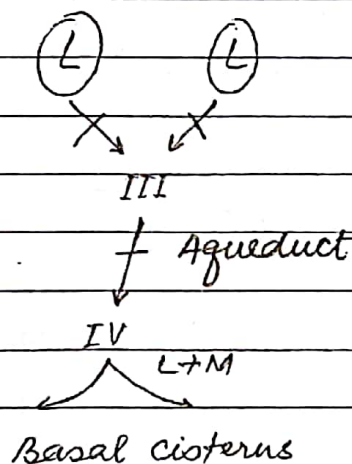
- Choroid plexus (75%) \rightarrow Lateral, III & IV ventricles.
- Extrachoroidal (25%) \rightarrow Capillary endothelium in brain parenchyma.

Rate of CSF production \rightarrow 20 ml/hr.

CSF volume in infants = 50 ml

adults = 150 ml.

CSF flow:



Obstructive/non-communicating.

- ① Aqueductal stenosis (M/C)
- ② Aqueductal gliosis
- ③ Arnold chiari Malformation

Downward displacement & hypoplasia in cerebellum leading to obliteration of cisterna magna.

Arnold-Chiari Type I - Adolescent/Adult

Type II - Newborn/ \bar{c} lumbosacral myelomeningocele.

④ Dandy-Walker Syndrome:



large cyst in posterior fossa communicating \bar{c} 4th ventricle.

- child has cerebellar hypoplasia



⑤ Vein of Galen malformation:

- M/c arteriovenous malformation in brain
- Sinus Venosus ASD.
- Obstructs aqueduct.
- Midline mass & dilated lateral ventricle.

Non-obstructive / communicating :

- Basal exudates

↳ TB

↳ Cryptococcal meningitis.

IOC for congenital Hydrocephalus — MRI.

Rx: Drugs → Furosemide

Acetazolamide.

VP Shunts → anastomose ~~large~~ ventricles to peritoneum.

↳ Gross hydrocephalus

↳ Thinned parenchyma.

↳ Complication — Blockage

Infection (Coagulase
-ve Staph).



Febrile Seizures:

- M/c seizure during childhood.
- Between 6 months - 5 yrs.

R/F for recurrence:

Major:

- Age < 1 yr
- Duration of fever < 24 hr.
- Fever $38 - 39^{\circ}\text{C}$ ($100.4 - 102.2^{\circ}\text{F}$)

Minor:

- Family H/O febrile seizures.
- Family H/O epilepsy.
- Complex febrile seizures.
- Daycare
- Male gender.
- Lower serum Na^{+} at time of presentation.

Having no risk factors carries a recurrence risk of approx 12%

- If 1 R/F $\rightarrow 25 - 50\%$
- 2 " $\rightarrow 50 - 59\%$
- ≥ 3 " $\rightarrow 73 - 100\%$

R/F	Risk for Subsequent Epilepsy
Simple febrile Seizure	1%
Recurrent " "	4%
Complex " "	6%
(> 15 min duration or recurrent \bar{c} in 24 hr)	
Fever < 1 before febrile seizure	11%
Family H/O epilepsy	18%
Neurodevelopmental abnormality (Mental Retardation).	33%



Complex febrile seizure complication:

- Prolonged febrile seizures.

MRI → Mesial temporal sclerosis

Temporal lobe + Hippocampal lobe.

Epilepsy in children:

Partial:

• Simple

• Complex - aura; automatisms.

Rx - Oxcarbamazepine &
Carbamazepine.

Ring Enhancing lesions.

Neurocysticercosis

- Solitary

- SCOLEX

Rx - Albendazole (DOC)



Before giving Albendazole

3-5 days of steroids given.

Tuberculoma.

- Large > 20mm

- Multiple

- Irregular margins

- Perilesional edema

- Midline shift.

Generalised epilepsy:

- Tonic

- Clonic

- GTCS [aura - GTCS] post ictal phase]



Drowsy, unconscious,
frothing, tongue bite
uprolling, incontinence.

- Atonic

- Myotonic



Rx - Sodium Valproate.



In < 2 yrs → It is hepatotoxic.

Absence Seizure:

- Blank stare < 30 sec.
- No aura/post ictal phase.
- Hyperventilation provokes.

Rx : Ethosuximide (DOC)
Valproate

Atypical Absence seizure:

- Myoclonic component

Rx - Valproate.

JME (Juvenile Myoclonic epilepsy):

- 12-18 yrs.
- Myoclonic jerks morning.
- Drops things.
- Gene & gene
- Family History.
- GTCs seizures 90%
- 1/3 Absence seizures.

EEG of JME → Generalised 4-6 Hz spike
+ photic stimulation.

Rx - Valproate (lifelong) → Excellent.



Infantile spasms / Salaam / West

- Flexor contractions of Head, trunk & extremities.
- 4-8 months
- EEG \rightarrow HYPSSARRHYTHMIA
 - \hookrightarrow Generalised chaotic high volume slow wave.
- Idiopathic / Cryptogenic \rightarrow Good.
- Secondary \rightarrow HIE, Structural malformation, Down's Syndrome, Tuberculous sclerosis.

Rx: - ACTH \rightarrow inhibit CRH. (DOC).

Vigabatrin \rightarrow In Tuberculous Sclerosis

STATUS EPILEPTICUS:

- Convulsion $>$ 30min

(OK)

Continuous 4w no regain of consciousness.

Vulnerable to hypoxia - Hippocampus, amygdala.
Thalamus, subcortical areas.

Rx: - i.v. Lorazepam (DOC) - longer $t_{1/2}$
0.05 mg/kg

- Midazolam + Phenytoin (20 mg/kg)
Repeat 10 mg/kg - 10 mg/kg
- Phenobarbitone 20 mg/kg - 10 mg/kg
- i.v. Valproate 20-30 mg/kg
- Midazolam infusion 2-20 mcg/kg/min.
- i.v. ~~levetiracetam~~ Levetiracetam 20 mg/kg.
- GA propofol; Thiopentone.



M/c/c of Status epilepticus → Febrile Seizures.



Rx → per rectal diazepam/
Buccal midazolam

Prevention of Febrile Seizures:

- No need
- Risk of Recurrence / concerned parents



Intermittent prophylaxis Oral CLOBAZAM / DIAZEPAM

↳ New BZD

for 48-72 hrs of fever.

MENINGITIS:

Cause:

	India	World
< 2 months	Klebsiella E. coli	Gr. B / D Streptococci E. coli
2 months - 3 yrs	H. influenzae type B	S. pneumoniae.
> 3 yrs	S. pneumoniae	S. pneumoniae Nisseria

Acute Bacterial meningitis:

- 95% cases occurs b/w 1 month to 5 yrs.
- Defect of complement system C5-C8 & properdin system
- meningococcal infection.
- T-lymphocyte defects (eg - AIDS / chemotherapy)
- Listeria monocytogenes / cryptococcus.



- congenital/acquired defects across mucocutaneous barrier → Pneumococci d/t cribriform plate.
- Lumbosacral meningocele & dermal sinus - Staph. & enteric bacteria.
- Penetrating CNS trauma / CSF shunt infection
 - coagulase -ve Staph.

Recurrent meningitis in CSF leak pt. M/c d/t pneumococcus.

Autosplenectomy (Sickle cell dysfunction/asplenia)
→ Pneumococcal infection
H. influenzae
Nisseria } capsulated.

Splenectomy vaccination time - 2 wks before.

Rx: Ceftriaxone (DOC)

DOC in Resistant pneumococci = Vancomycin + Ceftriaxone.

M/c Neurological sequel of meningitis:

- SNHL via aqueduct cochlear.
- Can we prevent SNHL?
 - Dexamethasone
 - 0.15 mg/kg
 - 30-60 mins before antibiotics.

Post exposure prophylaxis to contacts & doctors -

H. influenzae & Nisseria → i.m. Single dose ceftriaxone.

- Rifampicin X 2 days

Doctors → Fluoroquinolones.



Q. 3 yrs old diagnosed to have Hib meningitis. Ix done before discharge → BERA.
(Brainstem evoked response audiometry)

ENCEPHALITIS:

M/C/C = Enteroviruses (80% cases).

M/C sporadic = HSV-1

M/C aseptic meningitis in unimmunized children
— mumps.

Q. Child c fever & coma

Focal seizures.

CSF: Hemorrhagic

CT: Temporal hypodense; MRI → Hyperintense
— HSV-1 infection.

Localised temporal spike → HSV encephalitis.



DOC: i.v. Acyclovir.

Mortality rate of untreated herpes = 70%.

AFP (Acute Flaccid paralysis):

— Acute onset < 6 weeks

— < 15 yrs

— Rule out pseudoparalysis

↳ Septic arthritis

Osteomyelitis

• Scurvy; early syphilis

Hypokalaemia → Hypotonia.

— Asymmetrical AFP:

• Paralytic polio.



• Traumatic neuritis → i.m. injection (d/t)

- Symmetrical AFP:

• Transverse myelitis

↳ Herpes, Varicella, mycoplasma

Level → Thoracic area.

Rx: high dose i.v. Methyl prednisolone.

• Guillian Barre Syndrome (AIDP)

↳ Demyelinating.

- Diarrhea (By *Campylobacter jejuni*).
- Weakness occurs after 10 days.
- Areflexia (DTR absent)
- Symmetric
- Ascending → diaphragm involved.
- Plateaus → 4 wks.
- Sensory & autonomic changes.
- Also d/t Mycoplasma; salmonella; *S. pneumoniae*.

CSF	1st wk	2nd wk
Cells	10/4pf	10
Proteins	50mg/dl	500

→ Albuminocytological dissociation

- B/L symmetric demyelinating illness.

IOC for calcification: CT scan.

Rx: i.v. Ig (2g/kg)

↓ fails

Plasmapheresis.



Iv Ig indications:

- ↳ Kawasaki
- ↳ AIDP (Acute inflammatory Demyelinating Polyradiculopathy)
- ↳ Hypogammaglobulinemia
- ↳ Rh isoimmunisation.

General Pediatrics:

- Diarrhea
- ORS
- Zinc
- Malnutrition:

ORS:

Resomal	mmol/L	Old (WHO-ORS)	New/Universal
45	Sodium	90 (Cholera stool loss)	75 (Rotavirus 50-70)
40	Potassium	20	20
125	Glucose	111	75 (for facilitate diffusion of Na in cell)
70	Chloride	80	65
7	Citrate ⁻	10	10
+ Mn, Zn, Cu		311	245
300			

Low osmolarity ORS.

Citrate improves the self life of ORS.

Resomal → Rehydration solⁿ for malnutrition.

Hyperkalemia Hyponatremia.

Substrate concentration of components of ORS solⁿ:

NaCl — 2.6gms

KCl — 1.5gms

Trisodium citrate — 2.9gm

Glucose, anhydrous — 13.5gm



WHO dehydration:

- No
- Some \rightarrow Skin pinch slowly; thirsty
- Severe \rightarrow skin pinch very slowly; lethargic; oliguria.

R: No dehydration:

- Replace ongoing losses.
- 5-10 ml/kg.

Some dehydration:

- ORS 75 ml/kg over 4 hrs.

Severe dehydration:

- i.v. Ringer lactate 100 ml/kg.

	30 ml/kg	70 ml/kg
>1 yr	0.5 hr	2.5 hrs
<1 yr	1 hr	5 hrs.

WHO Zn in Acute Diarrhea:

- 2X RDA for 2 weeks.

Zn dose	RDA	Diarrhea
>6 months	10 mg	20 mg.
<6 months	5 mg	10 mg

Aerodermatitis an^ethropathica (periorificial dermatitis)

- d/t Zn deficiency.
- Nutritional
- Genetic (AR) \rightarrow Intestinal Zn transporter defect.
- Low Zn levels.



- Improve on Zn supplement.

Rx - Zn (3mg/kg) - elemental Zn.

Malnutrition:

	Marasmus	Kwashiorkor
Muscle Wasting	++	++
Edema		++
Hepatomegaly		++
Low albumin		++
Pigment		++
Appetite	Voracious	Poor
Sensorium	Alert	Lethargic

Weight/Age Criteria (Indian Academy of Pediatrics)

- Normal > 80% reference.
- Grade I → 71-80%
- Grade II → 61-70%
- Grade III → 51-60%
- Grade IV → ≤ 50%

for Edema ⇒ Add K

↳ Bad marker.

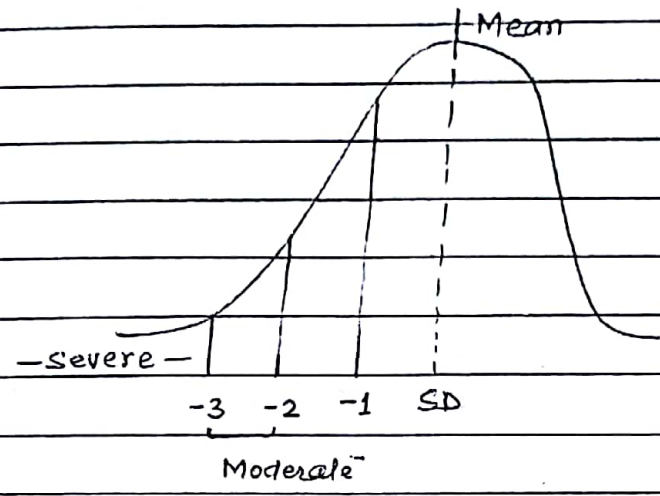
WHO

	Moderate	Severe
Weight/Height Acute (wasting)	71-79%	≤ 70
(N) > 80% reference		
Height/age Chronic (stunting)	86-89%	< 85%
(N) > 90% reference		



Symmetrical Edema

++++



Severe Acute Malnutrition:

Among children - 6-59 months of age.

Any of the following:

- ① Weight for height below -3 Standard deviation (SD or Z scores) of the median WHO growth references.
- ② Visible severe wasting.
- ③ Presence of bilateral edema.
- ④ Mid-arm circumference below 11.5 cm.

Below 6 months → Mid-arm circumference can't be used.

Criteria for admission - If child fails appetite test.

Criteria for passing Appetite test.

Body wt. (kg)	Min ^m amount of RUTF (Ready to use therapeutic factor) to be consumed for passing Appetite test (mL or grams)
< 4 kg	15 mL
4 - 6.9	25 mL



7-9.9

35 mL

10-14.9

50 mL

Mx:

Complications of SAM

Mx c in 1st 2 day c in 1st wk

S = Sugar (Blood Sugar $< 54 \text{ mg/dl}$ (OR) $< 3 \text{ mmol/L}$)
H = Hypothermia ($< 95.5^\circ\text{F}$ (OR) $< 35.5^\circ\text{C}$)
I = Infections (TB, Malaria, UTI)
EL = Electrolyte (Hypokalemia & Hyponatremia)
DE = Dehydration
D = Deficiency of Vitamins & minerals.

PHASE

STABILIZATION

REHABILITATION

Step

Day 1-2

Days 3-7

Weeks 2-6

1. Hypoglycemia _____
2. Hypothermia _____
3. Dehydration _____
4. Electrolytes _____
5. Infection _____
6. Micronutrients _____ No iron _____ c iron _____
7. Cautious feeding _____
8. Catch up growth. _____
9. Sensory stimulation _____
10. Prepare for follow up. _____

Feeding Rehabilitation: Cautious feeding

- F-75 containing 75 kcal/100ml & 0.9 g protein/100ml.

B = Begin feeds

E = Energy dense feeds.

S = Stimulation

T = Tender Love & Care



Days	Frequency	Vol/kg/feed	Vol/kg/day
1-2	2hrly	11 ml	130 ml
3-5	3hrly	16 ml	130 ml
6-7	4hrly	22 ml	130 ml

Energy dense feeds:

After the transition give:

- Frequent feeds (at least 4-hrly) of unlimited amounts of a catch-up formula.
- 150 - 220 kcal/kg/day.
- 4-6gm protein/kg/day.

Criteria for discharge:

- Weight for weight > 80% of Reference standard.
- Edema should be absent for 2wks.
- MAC > 12.5 cm.
- Weight gain > 5g/kg/day X 3 days.
- Appetite is good.
- Complete antibiotics.
- Care taker should have learnt, motivated.

Weight for age < Acute (Wasting)
Chronic (Stunting)

Age independent criteria — MAC (>12.5 cm)
b/w 1 to 5 yrs.

Age independent index — Kanawati & McLaren index
Rao & Singh index
Dugdale index
Quac stick index
Telford ratio



$$\# \text{ Osmolality} = 2 \times [\text{Na}] + [\text{Glucose}] / 18 + [\text{BUN}] / 2.8.$$

Pediatric Nephrology:

- Development
- Oliguria; polyuria
- Hematuria
- AKI
- CKD
- Nephrotic, Nephritic
- UTI

Topic

Development:

GFR:

- Newborn $\rightarrow 15 - 20 \text{ ml/min/1.73 m}^2$
- 3 months $\rightarrow 2/3^{\text{rd}}$ adult
- Like adults $\rightarrow 2 \text{ yrs of life.}$

- Tubular concⁿ:

- Adult morning urine osmolality $> 800 \text{ mosm/kg.}$
- Like adult 1yr.

- Nephrogenesis complete @ 36 wk of gestation.

- Barker's hypothesis:

preterm & IUGR \rightarrow hypertension in 2nd to 3rd decade
 \hookrightarrow Bcoz they have less no. of nephron.

- M/c asymptomatic abd. mass in 1-5 yrs - Wilms' tumour

- M/c abd. mass in newborn - Multicystic dysplastic kidney.

\downarrow
non-functional mass \rightarrow bunch of grapes
- 80% U/L.



OLIGURIA:

- U.O. $< 1 \text{ ml/kg/hr}$
- Common in AKI & Acute GN.

Causes of Non oliguric AKI — Aminoglycosides
— Neonatal renal failure.
— Resolving ATN (polyuria)

POLYURIA:

- U.O. $> 4\text{--}5 \text{ ml/kg/hr}$. [Polydipsia; Polyurea]

Approach —

- Blood glucose [RBS $> 200 \text{ mg/dl}$
or FBS $> 126 \text{ mg/dl}$]
↳ DM

- Venous Blood Gas.

(i) Hypokalemic hypochloric metabolic alkalosis
↓

Bartter Syndrome; Gitelman Syndrome.

Bartter Syndrome

- Severe; Early.
- Infancy
- Antenatal \rightarrow Polyhydramnios
 \rightarrow Nephrocalcinosis
- Hypercalciuria

Gitelman Syndrome

- Mild; Well preserved.
- Older child
- Hypo Mg
- Hypocalciuria.

(ii) Hyper Cl, hyper K, Normal anion gap
metabolic acidosis
↓

Renal tubular acidosis.

(iii) Normal $\left\{ \begin{array}{l} \text{Psychogenic} \\ \text{DI} \end{array} \right\}$ water deprivation
in psychogenic \rightarrow
Urine osmolality doubles



Q1 → Central/XLR Nephrogenic

- On Vasopressin challenge, urine osmolality doubles by 100% in central. Hence differentiated from XLR Nephrogenic.

Q 4yr; Polyuria & Polydipsia

Venous blood gas normal

Urine Osmolality

- Baseline 50 mOsm/kg.
- On water deprivation = 60 mOsm/kg
- On Vasopressin challenge = 70 mOsm/kg.

Δ = XLR Nephrogenic DI

DOC = Thiazides.

Best method of GFR estimation = Insulin clearance.

Schwartz eGFR formula:

$$= \frac{K \times \text{height in cm}}{\text{creatinine (mg/dl)}}$$

Formula depends on:

- Height, muscle mass.
- Method of estimation of creatinine.
[Jaffe's reaction].

S. creatinine is accurately measured by

- Enzyme assay / HPLC.



Value of k in Schwartz formula:

Low birth weight infant	—	0.33
Normal infant 0-18 months	—	0.45
Girls 2-16yrs	—	0.55
Boys 2-13yrs	—	0.55
Boys 13-16yrs	—	0.70

Schwartz method is independent of — Renal ~~function~~ function.

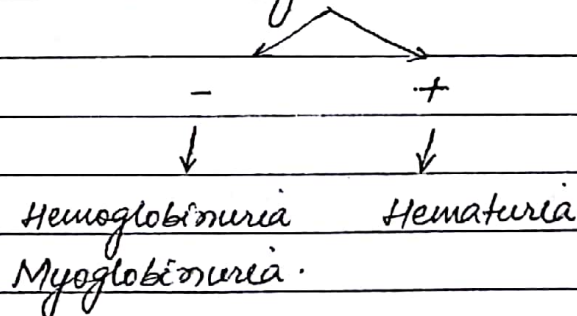
HEMATURIA:

Red urine:

- Beet root
- Phenolphthalein
- Rifampicin

Q. 5yrs old, Red urine. Urine 1-2 RBCs/hpf —
Intravascular hemolysis.

If Red urine → look for RBC's.



Hematuria:

- Gross
- Microscope → > 5 RBCs/hpf on centrifuged urine



	Glomerular	Extra glomerular
Cause —	- IgA ; MPGN; PSGN.	- Cystitis; Stones; Idiopathic hypercalciuria ↓ 24hr urine $\text{Ca}^{2+} > 4 \text{mg/kg}$.

Dysmorphic RBC ++

Colour	Cola	Bright Red.
Pain	Painless	Painful.

Protein in urine Proteinuria

Recurrent Gross Hematuria:

- IgA
- MPGN
- Idiopathic hypercalciuria.

PSGN doesn't recur.

Alport's Syndrome:

Triad:

Eye: Ant. Leucocoria

Ear: SNHL

Kidney: 75% Boys ESRD before <30yr.

- 80% X linked > AR 15% > AD 5%
- Collagen IV → α -5 domain (in G.B memb^r)

Good pasture Syndrome — α 3 domain of collagen IV abnormality.



INV: Electron Microscopy.

- ↳ Basement memb^r is irregular.
- ↳ Splitting of the lamina densa
- ↳ Lamellation
- ↳ Straited GBM
- ↳ Basket weave appearance (Classical)

Slit lamp examⁿ → Keratoconus
Lenticonus

Acute Kidney Injury (AKI):

Best Biomarker → ① Urine & NGAL

[Neutrophilic gelatinase.
associated lipocalin].

- ② Urine IL-18
- ③ Urine KIM-1 (Kidney injury molecule)
- ④ Urine L-FABP (Fatty acid binding protein).
- ⑤ Serum cystatin-C

Types:

- Prerenal
- Renal
- Post renal

PRE RENAL - Hypotension, Hypoxia

Newborns →; hypovolemia, burns, diarrhea.

RENAL - ATN > HUS

POST RENAL - Obstruction.

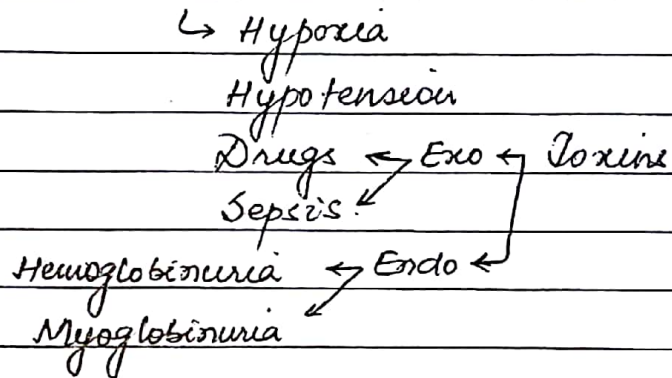


Indices	Pre renal	-Renal
Urinary Sodium (meq/L)	< 20	> 40
Urine Osmolality	> 500	< 300
B. urea/ Creatinine ratio	> 20:1	< 20:1
Fractional excretion of Na%	< 1	> 1
$= \frac{\text{Urine Na}}{\text{Serum Na}} \times \frac{\text{Serum Cr}}{\text{Urine Cr}}$		

Cause of AKI:

M/c/c in Children/Adults

Prerenal → ATN



Hemolytic Uremic Syndrome (HUS):

- Microangiopathic hemolytic anemia
- Thrombocytopenia
- AKI

- 90% follows diarrhea.
 - Developed: E. coli O157:H7.
 - Developing country: Shigella dysenteriae type I.
 - Germany June 2011: E. coli O104:H4
- SHIGA toxin → Cause Endothelial injury

↓
TMA (Thrombotic microangiopathy).

On PBS → Schistocytes are Δtic of HUS



Rx: ECULIZUMAB [DOC] → for PNH

↳ drug against Cs.

If not available → Plasmapheresis.

Complications:

↳ Insensible losses - 400ml/m²

- Fluid overload → Rx → Fluid restriction
- Hyperkalemia → cause arrhythmia / Sudden death.
- Dilutional hypo Na. Rx - Restrict fluid
- Dilutional anemia Rx - PRBCs if Hb < 6
- Metabolic acidosis.
- Hyper PO₄ → Hypo Ca
- HTN.

Hyperkalemia:

Rx - Glucose & insulin intravenously



C in 10-15 min.

- Ca²⁺ stabilises cardiac memb^r potential.

↳ only given in ECG changes in Hyperkalemia.



Calcium Gluconate iv.

~~Mechanism~~: Hyperkalemia Rx:

Transcellular shift into cell.

- Insulin C Dextrose.

- Nebulised salbutamol.

Cardioprotective → i.v. Calcium Gluconate.

↑ delivery Na to distal

• Furosemide

• i.v. NaHCO₃

Dialysis . .



K-binders \rightarrow Kaexylate polystyrene.

CKD (Chronic kidney disease):

Causes in children -

- < 5 yrs - Hypoplasia
Dysplasia
Posterior urethral valves (Boys)
- > 5 yrs - Acquired
 \rightarrow GN/HUS.

Features:

A = Azotemia

Acidosis (metabolic)

Anemia (Normocytic Normochromic)

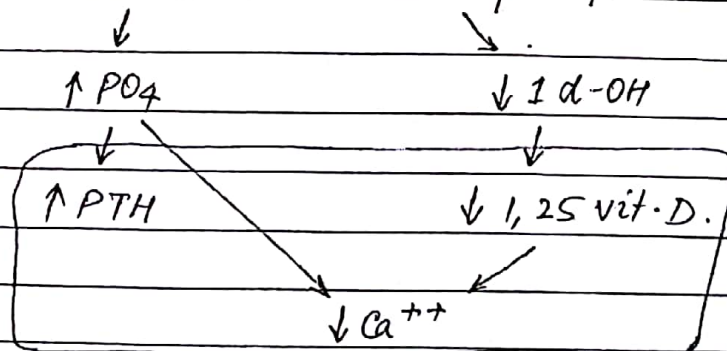
\rightarrow Rx - S/C rh EPO (Erythropoietin).

B = Bone ds

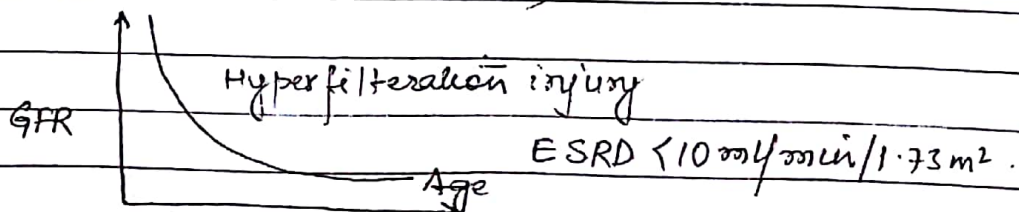
C = Cardiovascular complication \uparrow

G = Growth failure \leftarrow Multifactorial causes.

When GFR \downarrow - 40-60 mL/min/1.73 m²



Osteodystrophy
(CKD-MBD)





Nephrotic Syndrome:

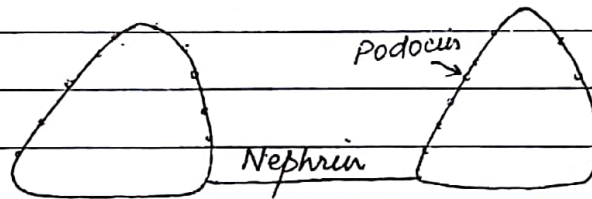
- Proteinuria $> 40 \text{ mg/m}^2/\text{hr}$ or $> 2 \text{ g/24hr}$
- Hypoproteinemia (Hypoalbuminemia)
- Hyperlipidemia (Cholesterol $> 200 \text{ mg/dL}$)
- Edema: S. Albumin $< 2.5 \text{ g/dL}$.

On light microscopy → Minimal changes.

On Electron " → Effacement of foot processes of podocytes.

DOC: Prednisolone

Cause of Edema in NS → Na^+ & H_2O reabsorption.



Gene	Protein	Disease
NPHS1 (chr 19)	Nephrin	Finnish Congenital nephrotic ($< 3 \text{ months}$) Rx: Nephrectomy.
NPHS2 (chr 1)	Podocin	Steroid resistant PSGS Rx: Calcineurin inhibitors ↳ Tacrolimus > Cyclosporine

Steroid toxic; Steroid dependent

- Cushingoid
- HTN
- Post. subcapsular cataract
- Impaired glucose tolerance
- Short

Rx - Oral cyclophosphamide for 12 wks.



UTI (Urinary Tract Infection):

Definition: Symptoms + Urine culture $>10^5$ CFU/ml

- Symptom - Fever
- E. coli
- Females [Boys <1 yr]
- Ascending [<1 yr \rightarrow Hematogenous]

Best urine specimen - Suprapubic aspiration.

- Asymptomatic bacteriuria \rightarrow Shouldn't be treated.
- M/c/c of UTI in children \rightarrow VUR
IOC for VUR \rightarrow MCU

VUR:

- Polar scarring \rightarrow DMSA nuclear scan
- Function \rightarrow MAG3 / DTPA nuclear scan.
- Prophylaxis \rightarrow Antibiotic of choice



Cotrimoxazole.

Sx \rightarrow Reimplantation of both ureter.

\hookrightarrow Indication:

- Break through UTIs
- Reterioration of Renal function.



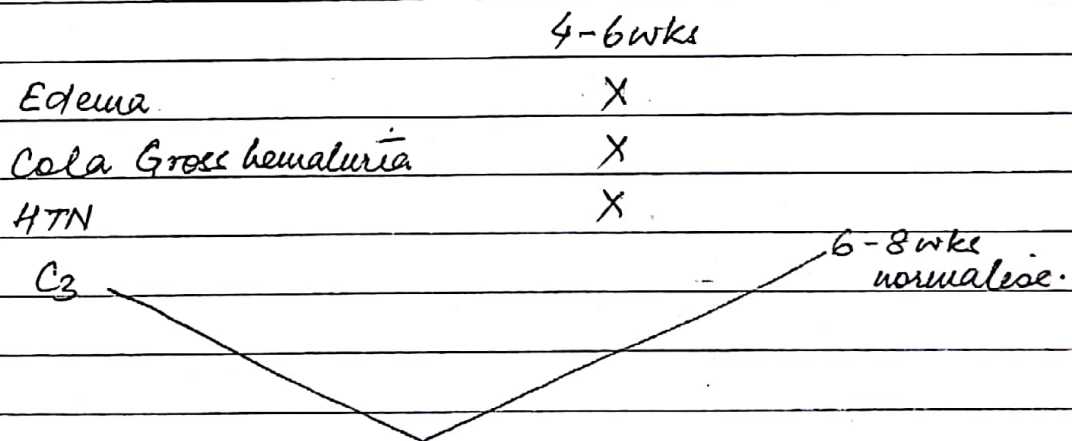
GLOMERULONEPHRITIS:

- Hematuria
- Edema
- HTN

M/c/c of GN → Post streptococcal GN.

Acute post streptococcal GN:

- Follows infection of throat (serotype 12) / skin (serotype 49) & nephritogenic strains of β -hemolytic streptococci.
- Age → 5-12 yrs
- Acute phase resolves in 4-6 wks but urine normalises in 1 yr.



In urine - Shows microscopic hematuria

↓
6-12 months to resolve.

Sore throat → Δ by ASO titre (1-2 wks)

Pyoderma → Δ by anti DNAase B (4-6 wks)

95% PSGN resolves

5% PSGN → CKD



Kidney Bx \rightarrow Endothelial & mesangial cell proliferation & obliteration of capillary lumen.
- Neutrophil infiltration

Immunofluorescence - Granular deposits of IgG & C3
"STARRY SKY"

Good pasture Syndrome has linear deposits of IgG & C3.

Subepithelial humps are ~~also~~ ^{also} ~~also~~ ^{also} of PSGN.

Q Persistently \downarrow C3 found in all except:

- a) Post streptococcal GN (normalise after 6-8 wks)
- b) Mesangiocapillary GN
- c) Cryoglobulinemia
- d) SLE
- e) IE
- f) Shunt nephritis
- g) Factor H-mutation \rightarrow HUS.